Natural History and Postoperative Evaluation of Complete Transposition of the Great Arteries

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In a total of 107 cases with transposition of the great arteries (TGA-s), 20 died preoperatively and 76 underwent surgical treatment: 11 palliative and 65 primary corrective surgeries, with hospital mortality 36.4% and 46.2% respectively. Of 31 survivors after Mustard operation, tricuspid regurgitant murmur and ECG abnormalities were recognized at 26.7% and 33.3% respectively in simple TGA, and 62.5% and 37.5% respectively in complicated TGA during an average follow-up of 4 years. Angiocardiogram, performed more than 2 years postoperatively, revealed decreased ejection fraction (EF) with compensatory increase of right ventricular end-diastolic volume (RVEDV): EF $0.43 \pm 0.04$, RVEDV $168 \pm 41$% of normal in simple TGA and $0.36 \pm 0.12$, $173 \pm 55$% in complicated TGA. In 3 long-term survivors of arterial switch operation (Stansel, Kaye), no serious complications were observed, and ejection fraction and echocardiographic findings of systemic ventricle showed an earlier normalizing than in Mustard operation: EF 0.65 or more when no VSD leakage and pre-ejection period (PEP)/ejection time (ET) $0.35 \pm 0.05$ after switch operation, $0.45 \pm 0.05$ in simple TGA and $0.47 \pm 0.07$ in complicated TGA after Mustard operation. From these long-term postoperative evaluations, our policy at present is to prefer arterial switch operation to Mustard operation in corrective surgery for TGA.

DEFINITELY ill cardiopulmonary condition of transposition of the great arteries (TGA) usually forces the patients to any of surgical treatments even in early infancy. Surgical treatments, however, are still in a developing field where more investigation and experience are needed both in atrial and arterial switch operations. This report describes our 10-year experience of mainly primary corrective surgeries for TGA, with special emphasis on long-term results of Mustard operation and arterial switch operation without relocation of coronary arteries (Stansel operation).

MATERIALS AND METHODS

From 1968 to 1979, final diagnosis of TGA was made on 107 patients in our institute either by cardiac catheterization with angiography (ACG), autopsy or corrective operation. Out of these 107 patients, 3 were excluded due to the associated complex anomalies which were considered as surgically uncorrectable and 8 were also excluded for the reason that they were still in the preoperative stage after examination. Thus, 96 patients constitute the present series.

Key Words:
- TGA
- Mustard operation
- Arterial switch operation
- Postoperative systemic ventricular function

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Patients were classified into the following three groups:
(1) TGA I; with intact ventricular septum,
(2) TGA II; with ventricular septal defect (VSD),
(3) TGA III; with VSD and pulmonary stenosis (PS).
Operation records and charts during admission and outpatient clinic were carefully analyzed regarding operative methods, results and postoperative course, especially the delayed occurrence of postoperative complications. Postoperative cardiac catheterization with ACG and echocardiography were performed at the time just before discharge, one to two months after operation, and they were carried out again more than two years after operation.
Postoperative follow-up periods ranged from 4 months to 9 years 2 months, longest in Mustard operation and shortest in Stansel operation.

RESULTS

1) Age and sex of each group
TGA I: 17 days to 9 years 8 months, 24 males and 6 females. TGA II: 23 days to 13 years 11 months, 38 males and 14 females. TGA III: 75 days to 15 years 2 months, 8 males and 6 females.

2) Natural history
Preoperative death was recognized in 20 patients (20.8% of 96 total patients): 5 out of 30 patients with TGA I, 12 out of 52 with TGA II and 3 out of 14 with TGA III (Fig. 1). In TGA I, excluding a 13 month-old girl who suffered from severe coagulation disturbance and unfortunately died at home while the operation was being deferred, the mean age of death was 45 days. In TGA II, it was 112 days excluding a 9 year-old boy, who had no chance of operation due to severe obstructive pulmonary vascular disease. In TGA III, the number of patients was small, but death seemed to occur according to the amount of pulmonary blood flow balancing the body development.

3) Operative results according to TGA types
Operation was performed on 76 patients: 11

<table>
<thead>
<tr>
<th>TABLE I OPERATIVE RESULTS OF TGA ACCORDING TO TYPES</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Palliative Op</strong></td>
</tr>
<tr>
<td>No. of pts</td>
</tr>
<tr>
<td>---------------</td>
</tr>
<tr>
<td>TGA I</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>TGA II</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>TGA III</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Total</td>
</tr>
<tr>
<td></td>
</tr>
</tbody>
</table>

(March '80, KAH)

palliative and 65 primary corrective surgeries (Table I). Palliative surgeries consisted of 6 Blalock-Taussig operations for TGA III, 2 Blalock-Hanlon operations for TGA I, 2 pulmonary arterial bandings (PAB) for TGA II and 1 Glenn anastomosis for TGA III. Hospital death occurred in 4 out of 11 cases and late death in 3, resulting in an overall mortality rate of 63.6%. Late death included 2 cases of asplenia syndrome. Primary corrective surgeries were carried out on 65 patients: 23 in TGA I (Mustard operation 21, Stansel operation 1 and Jatene operation 6,7,1), 38 in TGA II (Mustard operation 30, Stansel operation 5 and Jatene operation 3) and 4 in TGA III (Rastelli operation 8,9,2 and Daicoff operation 10,2). Hospital death was observed in 8 cases (34.8%) with TGA I, 20 cases (52.6%) with TGA II and 2 cases (50%) with TGA III, with the resultant overall hospital mortality rate of 46.2%. In each group, late death occurred in 1, 3 and 1 cases respectively, and the overall mortality rate was 14.3%.

4) Operative results according to methods

Operative results were analysed according to the operative methods (Table II). In 11 palliative operations, results were very unsatisfactory except for 6 Blalock-Taussig and 1 Glenn anastomoses in which no hospital deaths occurred. Blalock-Hanlon and PAB operations (2 cases respectively) were performed as emergency surgeries for intractable congestive heart failure and pulmonary dysfunction, but all died in early postoperative stage. Primary corrective surgeries were performed on 65 patients by 4 methods (Table II). Mustard operation was employed in 53 cases (81.5%). Arterial switch operation without relocation of coronary arteries (Stansel operation) was employed on 6 and anatomical correction (Jatene operation) on 4 patients, or 15.4% of 65 cases. Rastelli operation was carried out in 2 cases for total correction of TGA III. Operative results were poor for each method; mortality was 41.5% in Mustard operation and still higher in other procedures. As for the Mustard operation to TGA I, however, results have significantly improved during these past 3 years, resulting in no hospital deaths in 8 consecutive cases.

5) Long-term follow-up of Mustard operation

Postoperative long-term results after Mustard operation were analyzed clinically in 15 cases with TGA I and in 16 with TGA II (Fig. 2). Differentiation of tricuspid regurgitant (TR) murmur and that induced by residual VSD was made from the findings of postoperative ACG
and the appearance of a murmur previously not recognized. Positive ECG abnormalities included any ECG abnormalities, except sinus tachycardia, when they appeared even once in the postoperative follow-up period. Follow-up periods were from 9 months to 9 years 2 months (mean 4 years) in TGA I and from 6 months to 8 years 3 months (mean 4 years 5 months) in TGA II.

As for the postoperative medications, 11 cases (73.3%) have been followed up without medication in TGA I, in contrast to 8 cases (50%) in TGA II. Occurrence of TR murmur and ECG abnormalities are higher in TGA II than in TGA I: 62.5%, 37.5% in TGA II versus 26.7%, 33.3% in TGA I, respectively. No late death was observed in TGA I except for one case who died from hepatitis 9 months postoperatively. On the contrary, many problems have been recognized in postoperative courses in TGA II. Residual VSD was found in 7 cases, in which 2 died at 6 months and 10 months postoperatively from associated bacterial endocarditis. Sudden death occurred in 3 cases, whose postoperative periods were 1 year 7 months, 2 years 1 month and 5 years 10 months respectively. They all had TR murmur but only one case had ECG abnormality of paroxysmal atrial tachycardia.

6) Postoperative right ventricular function from ACG

Right ventricular end-diastolic volume (RVEDV) and ejection fraction (RVEF) in the patients who had undergone Mustard operation were analyzed with ACG in preoperative and postoperative stages (Fig. 3 and Fig. 4). There seems to be a gradual increase of RVEDV in the postoperative stage (in TGA I, 145% of normal at discharge and 168% at long-term stage; in TGA II, 169% and 173% of normal). On the other hand, RVEF decreased in both groups as time elapsed after operation though the value of decrease was small in TGA I (in TGA I, 0.47 at discharge and 0.43 at long-term stage; in TGA II, 0.43 and 0.36). These data suggest that decrease of RVEF is compensated by an increase of RVEDV to keep adequate cardiac output.

7) Arterial switch operation

There have been 10 arterial switch operations in our institute, mostly performed within the last year and a half (Table III). Cases consisted of 6 Stansel operations with 3 successes and 4 Jatene operations with no long-term survivors. Among these, however, it was concluded at a postoperative conference on dead cases that primary
Fig. 3. Right ventricular end-diastolic volume (RVEDV) before and after Mustard operation, obtained from angiocardiogram (ACG) at preoperative, early and late postoperative stage according to types of TGA.

Fig. 4. Right ventricular ejection fraction (RVEF) before and after Mustard operation, obtained from ACG at preoperative, early and late postoperative stage according to types of TGA. n = Number of patients. Mean value ± standard deviation.

switch operation was indicated erroneously in 3 cases (case No. 5, 6 and 7). Postoperative evaluation of 3 arterial switch operations (Stansel operation) was made by cardiac catheterization.
TABLE III  CASES OF ARTERIAL SWITCH OPERATION FOR TGA

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (mo-s)</th>
<th>BW (Kg)</th>
<th>Type</th>
<th>Assoc. anomalies</th>
<th>Operation</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>20</td>
<td>9.0</td>
<td>II</td>
<td>-</td>
<td>Jatene</td>
<td>Died, 8d, Renal failure</td>
</tr>
<tr>
<td>2</td>
<td>11</td>
<td>5.8</td>
<td>II</td>
<td>VSD IV, ASD, R-Ao, arch</td>
<td>Stansel</td>
<td>Excellent, 1y6m</td>
</tr>
<tr>
<td>3</td>
<td>13</td>
<td>7.0</td>
<td>II</td>
<td>ILL, PDA</td>
<td>Stansel</td>
<td>Died, 5d, PAT</td>
</tr>
<tr>
<td>4</td>
<td>11</td>
<td>6.2</td>
<td>II</td>
<td>PDA</td>
<td>Stansel</td>
<td>Good, 9m, Residual VSD</td>
</tr>
<tr>
<td>5</td>
<td>11</td>
<td>6.8</td>
<td>I</td>
<td>ASD</td>
<td>Stansel</td>
<td>Died, 1d, LV failure</td>
</tr>
<tr>
<td>6</td>
<td>16</td>
<td>7.4</td>
<td>II</td>
<td>TS severe, ASD PDA</td>
<td>Stansel</td>
<td>Died, 1d, LOS</td>
</tr>
<tr>
<td>7</td>
<td>4</td>
<td>5.6</td>
<td>I</td>
<td>ASD, PDA, PH</td>
<td>Jatene</td>
<td>Died, 1d, Resp. failure</td>
</tr>
<tr>
<td>8</td>
<td>10</td>
<td>7.2</td>
<td>II</td>
<td>R-Ao, arch</td>
<td>Jatene</td>
<td>Died, 2d, RV failure</td>
</tr>
<tr>
<td>9</td>
<td>14</td>
<td>8.0</td>
<td>II</td>
<td>PDA</td>
<td>Stansel</td>
<td>Excellent, 4m</td>
</tr>
<tr>
<td>10</td>
<td>3</td>
<td>3.8</td>
<td>II</td>
<td>PDA, Co/Ao</td>
<td>Jatene</td>
<td>Died, 3d, Resp. failure</td>
</tr>
</tbody>
</table>

(March '80, KAH)

BW = Body weight, VSD IV = Ventricular septal defect of muscular portion, ASD = Atrial septal defect, R-Ao. arch = Right aortic arch, ILL = Situs inversus, L-loop, L-position, PDA = Patent ductus arteriosus, TS = Tricuspid stenosis, PH = Pulmonary hypertension, Co/Ao = Coarctation of aorta, PAT = Paroxysmal atrial tachycardia, LV = Left ventricle, LOS = Low output syndrome, RV = Right ventricle.

TABLE IV  POSTOPERATIVE CLINICAL EVALUATION OF ARTERIAL SWITCH OPERATION (STANSEL OPERATION) FOR TGA.

<table>
<thead>
<tr>
<th>Case</th>
<th>Postop (mo-s)</th>
<th>RVEDV (% Normal)</th>
<th>RVEF</th>
<th>LVEF</th>
<th>Postop (mo-s)</th>
<th>CTR (%)</th>
<th>Clinical</th>
<th>ECG</th>
<th>Complication</th>
<th>Echo</th>
<th>PEP/ET</th>
<th>LV</th>
<th>RV</th>
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</thead>
<tbody>
<tr>
<td>2</td>
<td>2</td>
<td>144</td>
<td>0.42</td>
<td>0.65</td>
<td>18</td>
<td>68</td>
<td>NA</td>
<td>LVOL</td>
<td></td>
<td></td>
<td>0.29</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>2</td>
<td>184</td>
<td>0.28</td>
<td>0.49</td>
<td>9</td>
<td>65</td>
<td>LAD</td>
<td>CRBBB</td>
<td>VSD leak</td>
<td></td>
<td>0.33</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>1</td>
<td>163</td>
<td>0.45</td>
<td>0.71</td>
<td>4</td>
<td>67</td>
<td>RAD</td>
<td>CRBBB</td>
<td></td>
<td></td>
<td>0.38</td>
<td>0.48</td>
<td></td>
</tr>
</tbody>
</table>

(March '80, KAH)

ACG = Angiocardiogram, RVEDV = Right ventricular end-diastolic volume, RVEF = Right ventricular ejection fraction, LVEF = left ventricular ejection fraction, CTR = Cardio-thoracic ratio, NA = Normal axis, LVOL = Left ventricular overload, LAD = Left axis deviation, CRBBB = Complete right bundle branch block, RAD = Right axis deviation, VSD = Ventricular septal defect, PEP/ET = Pre-ejection period/ejection time, LV = Left ventricle, RV = Right ventricle.

with ACG at discharge and by clinical examinations, including echocardiogram, at the outpatient clinic (4 to 18 months postoperatively) (Table IV).

Ejection fractions of the systemic ventricle have normalized in the early postoperative stage (0.65 in case No. 2, 0.71 in case No. 9) except for a case of VSD leakage (0.49 in case No. 4). Clinical evaluation 4 months to one year and a half postoperatively revealed normalized cardiac function by echocardiogram (pre-ejection period/ejection time (PEP/ET) of systemic ventricle 0.29–0.38) and excellent physical and mental developments checked by pediatric specialists.

8) **Systemic ventricular function from echocardiogram after Mustard and Stansel operation**

Echocardiographic findings (PEP/ET) of systemic ventricle were compared among three groups: Mustard operation for TGA I and TGA II, and Stansel operation for TGA II (Fig. 5).
Fig.5. Comparison of Mustard operation with Stansel operation from postoperative echocardiographic findings of systemic ventricle.
PEP/ET = Pre-ejection period/ejection time, n = Number of patients, Mean value ± standard deviation.

CURRENT POLICY for TGA (Conclusion)

<table>
<thead>
<tr>
<th>Age</th>
<th>Neonatal</th>
<th>3 M</th>
<th>6 M</th>
<th>12 M</th>
<th>4 - 6 Y</th>
</tr>
</thead>
<tbody>
<tr>
<td>TGA I</td>
<td>BAS</td>
<td>Atrial Switch Op (Mustard, Senning)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Pulm. Art. Banding --- Arterial Switch Op (Jatene, Stansel)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>TGA II</td>
<td>(BAS)</td>
<td>Atrial Switch Op (Mustard, Senning)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Arterial Switch Op (Jatene, Stansel)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>TGA III</td>
<td>(BAS)</td>
<td>Systemic-Pulmonary Shunt Op</td>
<td>Rastelli Op (Blandes-Taussig)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

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Fig.6. Current policy of treatments for TGA.
BAS = Balloon atrial septostomy

Mustard operation for TGA I contained 11 patients, followed up from 2 years 1 month to 9 years 2 months (mean follow-up of 4 years 5 months) postoperatively and for TGA II, 6 patients from 4 years 10 months to 7 years 4 months (mean 5 years 7 months). Stansel operation contained 3 patients and 4 measurements, from 2 months to one year and a half after operation (mean follow-up of 8 months).

Systemic ventricular function after Stansel operation showed early normalizing (PEP/ET 0.35 ± 0.05), whereas it remained above normal after Mustard operation (in TGA I; 0.45 ± 0.05, in TGA II; 0.47 ± 0.07) with statistical significance between these two methods (p < 0.05).

COMMENTS
Though Senning\textsuperscript{11,12} and Mustard proce-
dures\textsuperscript{13} have proved successful in corrective surgeries for TGA, they have produced even more problems than they have solved.\textsuperscript{14–16} As postoperative complications of these atrial switch operations, arrhythmia\textsuperscript{17–19} systemic and pulmonic venous stenoses\textsuperscript{20,21} and tricuspid regurgitation\textsuperscript{22} have been the major obstacles to be overcome. To reduce these complications as much as possible, we have been using a fairly small sized pericardial baffle and excising the atrial septum thoroughly at the inflow portion of the superior vena cave into left atrium. In our series, except for one case in which reoperation was necessary for superior vena cave obstruction, so far no significant systemic and pulmonic venous stenoses has resulted without using patch enlargement of functional left atrium.

As for the postoperative ECG abnormality, although a rather high incidence was noted in this series, it becomes much lower if abnormalities are detected only from the last ECG tracings at the out-patient clinic (16.7% in TGA I, 30.8% in TGA II*). (\textsuperscript{\textsuperscript{*}}Reported by F. Okamoto at the 79th annual meeting of Japan surgical society at Sapporo, May 30th, 1979.) Myocardial temperature during operation has been checked continuously and topical cooling has been used as well as chemical cardiac arrest even under the condition of profound hypothermia.\textsuperscript{23} Even though recent technical improvement and myocardial protection have reduced those complications, there is inevitable problems when right ventricle is used as a systemic ventricle for long years.\textsuperscript{24} As shown in this report and elsewhere\textsuperscript{15,16} postoperative gradual increase in RVEDV and decrease in RVEF seem to suggest the catastrophic end-stage of right ventricular function represented by sudden death. This situation must be more likely in TGA II, where pulmonary vascular obstructive disease may progress earlier in the preoperative stage than in TGA I.\textsuperscript{25} And this seemed conversely proved by our investigation from the fact that in TGA II postoperative incidence of TR murmur and sudden death were significantly greater than in TGA I. On the other hand, operative success in arterial switch operations has still been very low in series reports possibly due to technical difficulties.\textsuperscript{5–7} Among those operations, arterial switch operation without relocation of coronary arteries (Stansel, Kaye) seemed to be easier than total anatomical correction (Jatene). Besides the advantages of preserving normal atrioventricular coordination and of avoiding arrhythmia and regurgitation of systemic atrioventricular valve, there is another advantage in this type of surgery, namely, that switching is possible in any anatomical condition of the coronary arteries. In young patients, however, this operation has the large disadvantage of using a valved conduit to reconstruct the main pulmonary artery. Although long-term results of the valved conduit are still controversial, we think a dysfunctioning valve could easily be exchanged as a conduit segment containing porcine valve. We have thus daringly used a valved conduit of large diameter (Hancock Model #150, 20 mm diameter) for those infants, so that exchanging only the valve portion would be enough even when the patient grows into an adult.

END REMARKS

From experiences of both atrial (Mustard) and arterial switch operations (Jatene, Stansel) and evaluation of long-term postoperative cardiac functions of both methods, we have arrived at the following directions for the treatment of TGA-s (Fig. 6):

1. For TGA I, after balloon atrial septostomy (BAS) at neonatal period, atrial switch operation (Mustard, Senning) should be performed during 3 to 12 months of age, if primary corrective surgery is scheduled. One may perform arterial switch operation (Jatene, Stansel) after training of left ventricle to pressure load by pulmonary arterial banding during early to middle infancy.\textsuperscript{26}

2. For TGA II, either atrial or arterial switching could be performed between mid to late infantile stage, but we positively prefer the latter from a consideration of long-term postoperative cardiac functions.

3. For TGA III, systemic to pulmonary shunt operation (Blalock-Taussig) at any infantile stage when necessary to increase pulmonary blood flow, and Rastelli operation at 4–6 years of age.

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