Physiologic Correction of Transposition of the Great Arteries

Indications for and Results of Operation in 32 Patients

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SUMMARY

The total management of a group of 32 patients with transposition of the great arteries (TGA) is reported. All but one had required creation of an atrial septal defect in infancy. Eighteen patients had no associated cardiac defects, and physiologic correction was performed between 8 months and 9% years of age. The only death in this group occurred 9 months after operation. Eight of nine patients with TGA and an associated ventricular septal defect (VSD) had required pulmonary artery banding in infancy. Physiologic correction was performed between 2% and 6 years of age. Three of these patients died during the postoperative period, and one died 9 months after operation. Two patients had TGA and pulmonary stenosis (PS). Physiologic correction was performed at 2 1/3 and 3% years of age. Both patients have done well. Three patients had TGA, VSD, and PS. Total correction was performed between 2 1/3 and 3% years of age. All three patients died in the postoperative period. The over-all operative mortality was 19%.

Our present plan of management has evolved from the experience reported above. All infants with TGA have balloon atrioseptostomy. If no VSD is present, physiologic correction is performed between 1 and 2 years of age. Patients with TGA and a VSD are catheterized at 8 months of age, and pulmonary banding is performed if pulmonary artery hypertension is present. Physiologic correction is performed at about 4 years of age. Patients with TGA, VSD, and PS may require systemic-pulmonary artery anastomosis. Correction at 5 years of age with the use of a homograft from the right ventricle to the pulmonary artery, as described by Rastelli and his associates, is recommended because of the poor results obtained with an intra-atrial baffle in this group.

Additional Indexing Words:
Congenital heart disease  Pulmonary artery banding  Balloon atrioseptostomy

TRANSPOSITION of the great arteries (TGA) is the most recent major form of congenital heart disease (CHD) to be corrected. The technical aspects of the operation have been described in detail. However, the total management of this complex group of patients, including the precise indications for operation, is still not firmly established. The present report reviews our experience with the operative management of 32 patients with TGA in order to determine these factors.

Clinical Material

Thirty-two patients ranging in age from 8 months to 9 years, had physiologic correction of TGA by a modification of the intra-atrial baffle technic described by Mustard. All patients were studied by cardiac catheterization and cine-angiography early in infancy. Since May, 1965, each patient has had balloon atrioseptostomy.
### Table 1

**Simple Transposition of the Great Arteries: Patient Information and Physiologic Correction of TGA**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr)</th>
<th>Weight (kg)</th>
<th>BAS</th>
<th>Preoperative studies</th>
<th>Discharge studies</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>M.C.</td>
<td>2 9/12</td>
<td>11.0</td>
<td>30 days</td>
<td>11.0 36</td>
<td>— —</td>
<td>Hb</td>
</tr>
<tr>
<td>C.S.</td>
<td>2 9/12</td>
<td>10.0</td>
<td>4 days</td>
<td>15.0 25 35/8</td>
<td>— —</td>
<td>210‡</td>
</tr>
<tr>
<td>D.M.S.</td>
<td>3 7/12</td>
<td>8.4</td>
<td>11 months (B-H)</td>
<td>15.6 42 25/5 15/8</td>
<td>50 NSR</td>
<td>Postoperative bleeding, pneumonia, CVA, GI bleeding, tracheostomy</td>
</tr>
<tr>
<td>R.R.</td>
<td>2 4/12</td>
<td>10.2</td>
<td>42 days</td>
<td>17.0 40 32/2 20/7</td>
<td>45 Nodal</td>
<td>—</td>
</tr>
<tr>
<td>K.S.</td>
<td>2 5/12</td>
<td>11.5</td>
<td>9 days</td>
<td>12.9 26 45/2 45/12</td>
<td>230§ NSR</td>
<td>Respiratory failure, tracheostomy</td>
</tr>
<tr>
<td>D.S.</td>
<td>2 1/12</td>
<td>5.3</td>
<td>8 days</td>
<td>11.0 23 33/3 36/20</td>
<td>60 Nodal</td>
<td>Postoperative septicemia, tracheostomy</td>
</tr>
<tr>
<td>A.F.</td>
<td>7 4/12</td>
<td>22.5</td>
<td>60 days (B-H)</td>
<td>22.4 37† 24/0</td>
<td>42 NSR</td>
<td>—</td>
</tr>
<tr>
<td>K.E.</td>
<td>2 1/12</td>
<td>9.3</td>
<td>1 day</td>
<td>18.0 25 40/2 24/10</td>
<td>43 NSR</td>
<td>Preoperative CVA, postoperative low cardiac output</td>
</tr>
<tr>
<td>K.D.E.</td>
<td>2</td>
<td>9.2</td>
<td>74 days</td>
<td>19.0 25 62/5 55/32</td>
<td>46 Nodal</td>
<td>Respiratory failure, tracheostomy</td>
</tr>
<tr>
<td>G.G.</td>
<td>2 6/12</td>
<td>5.7</td>
<td>5 days</td>
<td>14.3 20 22/4 15/8</td>
<td>61 NSR</td>
<td>—</td>
</tr>
<tr>
<td>J.I.</td>
<td>2 4/12</td>
<td>11.0</td>
<td>20 days</td>
<td>16.8 37 27/3 19/12</td>
<td>53 Nodal</td>
<td>—</td>
</tr>
<tr>
<td>S.H.</td>
<td>1 8/12</td>
<td>8.7</td>
<td>4 days</td>
<td>15.9 25 30/6 15/8</td>
<td>98§ NSR</td>
<td>Postoperative bleeding</td>
</tr>
<tr>
<td>S.L.</td>
<td>1 4/12</td>
<td>7.5</td>
<td>22 days</td>
<td>11.0 22 50/11 47/17</td>
<td>50 NSR</td>
<td>Two CVA preoperatively</td>
</tr>
<tr>
<td>C.E.</td>
<td>2 6/12</td>
<td>12.8</td>
<td>1 day</td>
<td>20.0 35 35/3 15/5</td>
<td>75 NSR</td>
<td>No coronary sinus present</td>
</tr>
<tr>
<td>S.W.</td>
<td>8/12</td>
<td>8.0</td>
<td>34 days</td>
<td>17.4 32 30/4 15/2</td>
<td>60 NSR</td>
<td>—</td>
</tr>
<tr>
<td>G.W.</td>
<td>4 11/12</td>
<td>13.3</td>
<td>2 days</td>
<td>17.0 40 13/11 27/7</td>
<td>63‡ Nodal</td>
<td>PAB performed in infancy, postoperative CVA</td>
</tr>
<tr>
<td>F.B.</td>
<td>9 5/12</td>
<td>22.2</td>
<td>80 days (open septectomy)</td>
<td>11.5 37 80/7 40/20</td>
<td>40 NSR</td>
<td>PAB performed in infancy, PDA ligated at time of correction, postoperative bleeding</td>
</tr>
<tr>
<td>W.M.</td>
<td>2 3/12</td>
<td>8.8</td>
<td>33 days</td>
<td>16.8 28 38/4 19/9</td>
<td>50 NSR</td>
<td>Preoperative CVA</td>
</tr>
</tbody>
</table>

*Key: BAS = balloon atrioseptostomy; Hb = hemoglobin (g/100ml); PaO₂ = arterial oxygen tension (mm Hg), fraction inspired oxygen (fiO₂) of 0.21; LV = left ventricular pressure (mm Hg); PA = pulmonary artery pressure (mm Hg); B-H = Blalock-Hanlon operation; NSR = normal sinus rhythm; CVA = cerebrovascular accident; GI = gastrointestinal; PAB = pulmonary artery banding; PDA = patent ductus arteriosus.

†Fraction inspired oxygen (fiO₂) of 0.4.

‡fiO₂ of 0.6.

§fiO₂ of 0.1.
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**Table 2**

**Complex Transposition of the Great Arteries: Patient Information and Physiologic Correction of TGA**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr)</th>
<th>Weight (kg)</th>
<th>BAS</th>
<th>Associated defects</th>
<th>Previous operative procedures</th>
<th>Preoperative studies</th>
</tr>
</thead>
<tbody>
<tr>
<td>W.M.</td>
<td>6</td>
<td>19.0</td>
<td>10 months (B-H)</td>
<td>VSD</td>
<td>PAB 10 months</td>
<td>21.0 39 90/0 12/5</td>
</tr>
<tr>
<td>D.P.</td>
<td>3 3/12</td>
<td>12.0</td>
<td>7 days</td>
<td>VSD</td>
<td>PAB 18 months</td>
<td>17.4 34 85/6 42/12</td>
</tr>
<tr>
<td>B.W.</td>
<td>3 1/12</td>
<td>12.0</td>
<td>66 days</td>
<td>VSD</td>
<td>PAB 20 months</td>
<td>16.0 40 135/12 18/12</td>
</tr>
<tr>
<td>S.Y.</td>
<td>3 5/12</td>
<td>15.0</td>
<td>1 day, 18 months (B-H)</td>
<td>VSD</td>
<td>PAB 18 months</td>
<td>20.0 38 77/10 35/25</td>
</tr>
<tr>
<td>S.P.</td>
<td>3 6/12</td>
<td>9.9</td>
<td>21 days</td>
<td>VSD</td>
<td>None</td>
<td>16.5 37 35/3 35/18</td>
</tr>
<tr>
<td>J.H.</td>
<td>2 9/12</td>
<td>12.1</td>
<td>40 days (B-H)</td>
<td>VSD</td>
<td>PAB 6 weeks</td>
<td>21.0 29 90/4 30/8</td>
</tr>
<tr>
<td>T.R.</td>
<td>5 6/12</td>
<td>18.0</td>
<td>No</td>
<td>Taussig-Bing</td>
<td>PAB 4 5/12 yr</td>
<td>22.6 29 95/5 55/37</td>
</tr>
<tr>
<td>J.C.</td>
<td>2 3/12</td>
<td>10.0</td>
<td>30 days</td>
<td>Taussig-Bing</td>
<td>PAB 6 weeks</td>
<td>16.0 30 95/6 17/13</td>
</tr>
<tr>
<td>C.F.</td>
<td>2 4/12</td>
<td>9.7</td>
<td>28 days</td>
<td>VSD, PDA, aortic arch atresia</td>
<td>PAB, ligation PDA, aortoplasty, arch reconstruction 10 months</td>
<td>19.0 47† 122/7 65/35</td>
</tr>
<tr>
<td>M.P.</td>
<td>3 9/12</td>
<td>11.6</td>
<td>17 months</td>
<td>PS</td>
<td>None</td>
<td>18.0 25 98/12 10/8</td>
</tr>
<tr>
<td>J.S.</td>
<td>2 4/12</td>
<td>10.0</td>
<td>4 days</td>
<td>PS</td>
<td>Ascending aorta-right PA anastomosis 18 months</td>
<td>16.6 29 100/5 17/8</td>
</tr>
<tr>
<td>G.B.</td>
<td>2 5/12</td>
<td>8.2</td>
<td>1 day, 2 yr</td>
<td>VSD, PS</td>
<td>None</td>
<td>22.0 29 72/2 44/12</td>
</tr>
<tr>
<td>M.S.</td>
<td>2 11/12</td>
<td>11.6</td>
<td>12 days</td>
<td>VSD, PS</td>
<td>None</td>
<td>22.8 29 85/0 24/14</td>
</tr>
<tr>
<td>G.R.</td>
<td>3 6/12</td>
<td>11.6</td>
<td>76 days</td>
<td>VSD, peripheral PS</td>
<td>None</td>
<td>14.3 35 90/10 18/10</td>
</tr>
</tbody>
</table>

*Key: BAS = balloon atrioseptostomy; Hb = hemoglobin (g/100 ml); PaO₂ = arterial oxygen tension (mm Hg), fraction inspired oxygen (fiO₂) of 0.21; LV = left ventricular pressure (mm Hg); PA = pulmonary artery pressure (mm Hg); B-H = Blalock-Hanlon operation; VSD = ventricular septal defect; PAB = pulmonary artery banding; NSR = normal sinus rhythm; PDA = patent ductus arteriosus; PS = pulmonic stenosis; CVA = cerebrovascular accident.

†Fraction inspired oxygen (fiO₂) of 0.6.

(BAS) at the time of the initial catheterization (25 of the 32 patients). Repeat studies were performed in any patient who did poorly or who required pulmonary artery banding and in all patients prior to total correction.

**Group 1: Simple Transposition**

Patients with simple TGA were those without a ventricular septal defect (VSD) or without significant pulmonary outflow obstruction (left ventricle to pulmonary artery gradient less than 20 mm Hg). One patient in this group did have a small patent ductus arteriosus (PDA). This group of 18 patients ranged in age from 8/10 to 95/12 years, although the majority were about 2 years old. Their average weight was 10.8 kg. Detailed patient information on this group is presented in table 1. Fourteen had a BAS early in infancy, while four had surgical excision of the atrial septum by the Blalock-Hanlon technic or during inflow occlusion prior to the development of the BAS technic. Two infants.
### Suture closure VSD (12 mm), reconstruct PA
- Discharge studies: 
  - \(\text{PaO}_2\): —
  - Rhythm: —
- Remarks: Died 12 hours postoperatively of concealed bleeding and cardiac tamponade.

### Suture closure VSD (8 mm), removal PAB
- Discharge studies: 
  - \(\text{PaO}_2\): 205†
  - Rhythm: Atrial flutter
- Remarks: —

### Suture closure VSD (10 mm), removal PAB
- Discharge studies: 
  - \(\text{PaO}_2\): 52
  - Rhythm: NSR
- Remarks: Reoperation for postoperative bleeding. Edema seen at follow-up examination. Cardiac cath 5 months postoperative PA = 90/50(60), marked systemic atrioventricular valve insufficiency present. Has cleared with medical therapy.

### Suture closure VSD (10 mm)
- Discharge studies: 
  - \(\text{PaO}_2\): 48
  - Rhythm: NSR
- Remarks: —

### Patch closure of VSD (10 mm)
- Discharge studies: 
  - \(\text{PaO}_2\): 165†
  - Rhythm: NSR
- Remarks: Visceral larva migrans.

### Patch closure of VSD, reconstruct PA
- Discharge studies: 
  - \(\text{PaO}_2\): —
  - Rhythm: —
- Remarks: Low output syndrome, died 48 hours postoperatively. No anatomic cause found for death.

### Contour patch closure VSD, removal PAB
- Discharge studies: 
  - \(\text{PaO}_2\): 50
  - Rhythm: Varying P-R interval
- Remarks: Died at home of overwhelming viral infection 4 months postoperatively.

### Contour patch closure VSD, reconstruct PA
- Discharge studies: 
  - \(\text{PaO}_2\): —
  - Rhythm: —
- Remarks: Died 15 hours postoperatively with low output syndrome. Autopsy showed repair to be intact.

### Suture closure VSD (10 mm), removal of PAB
- Discharge studies: 
  - \(\text{PaO}_2\): 60
  - Rhythm: ?

<table>
<thead>
<tr>
<th>Procedure</th>
<th>(\text{PaO}_2)</th>
<th>Rhythm</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Suture closure VSD (12 mm), reconstruct PA</td>
<td>—</td>
<td>—</td>
<td>Died 12 hours postoperatively of concealed bleeding and cardiac tamponade</td>
</tr>
<tr>
<td>Suture closure VSD (8 mm), removal PAB</td>
<td>205†</td>
<td>Atrial flutter</td>
<td>—</td>
</tr>
<tr>
<td>Suture closure VSD (10 mm), removal PAB</td>
<td>52</td>
<td>NSR</td>
<td>Reoperation for postoperative bleeding. Edema seen at follow-up examination. Cardiac cath 5 months postoperative PA = 90/50(60), marked systemic atrioventricular valve insufficiency present. Has cleared with medical therapy.</td>
</tr>
<tr>
<td>Suture closure VSD (10 mm)</td>
<td>48</td>
<td>NSR</td>
<td>—</td>
</tr>
<tr>
<td>Patch closure of VSD (10 mm)</td>
<td>165†</td>
<td>NSR</td>
<td>Visceral larva migrans.</td>
</tr>
<tr>
<td>Patch closure of VSD, reconstruct PA</td>
<td>—</td>
<td>—</td>
<td>Low output syndrome, died 48 hours postoperatively. No anatomic cause found for death.</td>
</tr>
<tr>
<td>Contour patch closure VSD, removal PAB</td>
<td>50</td>
<td>Varying P-R interval</td>
<td>Died at home of overwhelming viral infection 4 months postoperatively.</td>
</tr>
<tr>
<td>Contour patch closure VSD, reconstruct PA</td>
<td>—</td>
<td>—</td>
<td>Died 15 hours postoperatively with low output syndrome. Autopsy showed repair to be intact.</td>
</tr>
<tr>
<td>Suture closure VSD (10 mm), removal of PAB</td>
<td>60</td>
<td>?</td>
<td>Tracheostomy. Has residual coarctation of the aorta. One episode of congestive heart failure 2 months after operation.</td>
</tr>
</tbody>
</table>

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early in this series had a pulmonary artery banding (PAB) at the time of the septectomy, even though a VSD was not present. Three of the patients had cerebrovascular accidents (CVA) during the interim between the BAS and their corrective operation.

Indications for corrective operation were based on age and weight. Aside from the older children the majority of patients in this group were operated on when they attained a weight of 10 kg. In two infants no significant weight gain occurred after they attained 5 kg, and correction was performed at that weight.

Total correction was performed by means of an intra-atrial pericardial baffle was previously described. In the patient with a patent ductus arteriosus and pulmonary artery banding, the ductus was ligated at the time of physiologic correction through the median sternotomy as described by Kirklin. The Teflon fabric PAB was removed without difficulty, and no pulmonary artery reconstruction was required. In one patient with a cotton fabric PAB, the pulmonary artery was reconstructed with a pericardial patch graft. In the last eight patients of this group, the new left atrium was enlarged with a large (2.5 by 2.5 cm) diamond-shaped pericardial patch, as previously described, in order to prevent pulmonary venous obstruction.

### Group II: Complex Transposition

This group includes all patients with a VSD...
or significant pulmonary stenosis (PS), or both, in addition to the transposition. Detailed patient information on this group of 14 patients is presented in table 2.

A. Transposition and Ventricular Septal Defect

There were nine patients in this group, ranging in age from 2 3/12 to 6 years. The majority were about 3 years of age, and their mean weight was 13.5 kg (table 2). Each patient had a large VSD (8 mm in diameter or greater), and all but one had pulmonary artery hypertension (PAH) with pulmonary to systemic pressure ratios greater than 0.70. Four patients had pressure ratios greater than 0.95. Two patients in this group had Taussig-Bing transposition with a subpulmonic VSD and an overriding pulmonary artery. The remainder had membranous septal defects which in two instances extended into the endocardial cushion area. One patient also had a preductal coarctation with a large patent ductus arteriosus and a hypoplastic aortic arch.

Balloon atrioseptostomy was performed in six of these patients early in infancy. One of them subsequently required operative septectomy. Two additional patients had a blalock-Hanlon operation. One patient with Taussig-Bing transposition did not have his atrial septum opened. Pulmonary artery banding was performed in all eight patients with PAH. One patient with Taussig-Bing transposition required an emergency PAB 2 weeks after BAS. Two had PAB at the time of the Blalock-Hanlon operation and five more, including the older patient with Taussig-Bing transposition, had the banding performed electively between 10 and 20 months of age for persistent PAH. Aortoplasty, reconstruction of the hypoplastic aortic arch, and ligation of the PDA were performed in the patient with a preductal coarctation. All patients did well following PAB. The patient with the preductal coarctation sustained a CVA 1 week after the banding which may have been related to an indwelling venous catheter. Although only one patient had dyspnea on exertion after PAB four of the eight had a hemoglobin level of about 20 g/100 ml prior to total correction.

Indication for corrective operation in this group was attainment of a weight above 12 kg. Several patients were operated on at a lower weight because they had stopped gaining. A median sternotomy was employed. The pulmonary artery band could almost always be removed by simple transection if it was Teflon fabric. Reconstruction of the pulmonary artery with a pericardial patch graft was required in the two patients with cotton fabric bands. Anoxic arrest was employed during VSD closure. The defect was exposed through the atrium by retracting the tricuspid valve and overlying chordae tendineae. In five patients the defect was closed with several interrupted mattress sutures tied over Teflon felt pledges, while in two others the defect was closed with a woven Dacron patch. In the two patients with Taussig-Bing transposition, the defect was closed with a Dacron patch positioned in such a manner that the pulmonary artery arose from the left ventricle. All patients then had placement of an intra-atrial baffle and enlargement of the physiologic left atrium by a diamond-shaped pericardial patch.

B. Transposition and Pulmonic Stenosis

The two patients in this group had severe subvalvular pulmonic stenosis and underwent BAS in infancy (table 2). One infant became progressively more cyanotic and dyspneic and, at 18 months of age, had an ascending aorta-right pulmonary artery anastomosis. He tolerated this procedure well but remained markedly cyanotic.

Increasing cyanosis was the indication for physiologic correction in these patients. Operation was performed through a median sternotomy. The pulmonic obstruction was visualized by opening the pulmonary artery and retracting the pulmonary valve leaflets. A subvalvular band of fibrous tissue and muscle could be seen in each instance. This was excised along the anterolateral margin so as not to injure the mitral valve or the conduction system in the ventricular septum. An intra-atrial pericardial baffle was then sutured into place. In one patient an ascending aorta-right pulmonary artery anastomosis 2 mm in diameter was readily closed through the ascending aorta just after the initiation of cardiopulmonary bypass.

C. Transposition, Ventricular Septal Defect, and Pulmonic Stenosis

The three patients in this group averaged 3 years of age (table 2). BAS had been performed in infancy in each patient. Two patients had marked cyanosis and a rising hematocrit; one subsequently had a CVA. This latter patient was found to have only a very small atrial septal defect (7 mm). BAS had been performed elsewhere. All three children had correction of TGA as described above. Each patient in this group had a large VSD measuring 10 to 12 mm in diameter. Two were retrocristal defects while one was in the subaortic location. In two patients the VSD was closed with a Dacron patch while in one patient direct suture was employed. The subvalvular obstruction in two patients was resected through to main PA as in those with TGA and PS. The pulmonary stenosis in the third patient was in the branch pulmonary arteries. Operative correction of branch stenosis was not
physiologic correction of transposition

performed because, after closure of the VSD and insertion of the intra-atrial pericardial baffle, left ventricular systolic pressure was only 40 mm Hg when the aortic systolic pressure was 80 mm Hg at a time when the cardiac output appeared adequate.

All patients had continuous, direct arterial pressure, central venous pressure, and ECG monitoring for a minimum of 48 hours after operation. Arterial pH, PaO₂, and PaCO₂, as well as serum sodium and potassium determinations, were performed frequently. All patients had cardiac output determinations performed in the early postoperative period and for 1 to 3 days thereafter with a dye-dilution technic.

Patients with adequate respiratory function, as manifested by physiologic and clinical criteria, were extubated within 24 hours after operation. In five patients tracheostomy was performed in the postoperative period since prolonged mechanical ventilation was required.

Results

Twenty-six of the 32 patients with TGA survived the postoperative period, resulting in an operative mortality of 19% (table 3).

Group I

All 18 patients with isolated TGA survived the postoperative period. Eleven of these were previously reported in detail. Although many of the patients had arterial oxygen tension below normal at the time of discharge, a gradual increase in this value has been observed. One patient operated on early in our experience died 9 months after operation and was found at autopsy to have a contracted pericardial baffle which had caused severe pulmonary venous hypertension. In addition, partial anomalous pulmonary venous drainage had been produced by failure to include all four pulmonary veins in the new pulmonary venous atrium. The other 17 patients in this group have shown good growth and development since hospital discharge.

The complications in the first 11 isolated TGA have previously been described. Since introduction of the technic for enlargement of the new left atrium, further pulmonary complications have not been encountered in this group. Postoperative bleeding was the most common complication and occurred in three of the 18 patients. Two patients had a CVA of unknown causes in the early postoperative period but made full recovery. The three patients who sustained a CVA prior to total correction have also made an excellent recovery with little residual neurologic deficit. Half of the patients in this group had a nodal rhythm immediately after operation, but only two required transient cardiac pacing. Six patients had a nodal rhythm at a rate of about 100 beats/min at the time of discharge which persisted in some instances as long as 7 months after operation. The remainder had an atrial rhythm at discharge.

Group II A

Three of the nine patients with TGA and VSD died in the postoperative period. One died of unrecognized hemorrhage and cardiac tamponade 12 hours after operation, while two died of the low cardiac output syndrome 15 and 48 hours after operation. One patient was reoperated on for postoperative bleeding but subsequently did well. Pulmonary complications were surprisingly uncommon in this group in spite of the fact that eight of nine had had PAH prior to their previous PAB. However, four patients did require postoperative mechanical ventilation. Three of the nine patients had slow nodal rhythms after operation and required cardiac pacing. All six survivors had an atrial rhythm at discharge; one has atrial flutter controlled with digitalis.

Of the remaining six patients, one with a Taussig-Bing transposition died at home 4 months after operation, apparently of viral septicemia, following a brief febrile illness of

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Patients</th>
<th>Operative deaths (30 days)</th>
<th>Late deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>TGA – simple</td>
<td>18</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>TGA – complex</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>VSD</td>
<td>9</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>PS</td>
<td>2</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>VSD-PS</td>
<td>3</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>32</td>
<td>6</td>
<td>2</td>
</tr>
</tbody>
</table>

*See text for abbreviations.
less than 8 hours. Autopsy showed an intact cardiac repair and no apparent cause of death. Two patients developed congestive heart failure some months after total correction. One of these patients has a residual coarctation of the aorta. The other patient was recatheterized 5 months after operation to determine the etiology of the congestive heart failure and was found to have severe PAH (mean pulmonary to systemic pressure ratio 0.75) secondary to systemic atrioventricular (tricuspid) valve insufficiency and subsequent pulmonary venous hypertension. He has responded to medical management and now has minimal tricuspid insufficiency and no evidence of pulmonary hypertension.

**Group II B**

Both patients with transposition and pulmonary outflow tract obstruction have done well 10 and 12 months since discharge. One had a transient CVA 48 hours after operation but now has no residual neurologic deficit. Both patients have a residual PS murmur, but they have grown well and are active children. Both patients are in normal sinus rhythm.

**Group II C**

All three patients with TGA, VSD, and PS died in the immediate postoperative period. One died suddenly 48 hours following operation after an initially uncomplicated course. At autopsy the repair was intact, and no cause of death could be determined. This patient required postoperative cardiac pacing for a nodal bradycardia, and pacemaker failure could not be excluded. Each of the other two patients sustained a CVA of unknown cause early in the postoperative period and died 48 and 72 hours after operation. Both had severe pulmonary problems in the postoperative period.

One patient (G.B.) had postoperative respiratory failure and required mechanical ventilation. At autopsy a subarachnoid hemorrhage was present with areas of old and recent brain infarction. Severe pulmonary vascular disease involving both the intima and media was present.

The remaining patient (G.R.) had completeatelectasis of one lung in the postoperative period; severe pulmonary edema of the opposite lung followed, and mechanical ventilation was required. Cardiac pacing was employed because of a nodal bradycardia. A subarachnoid hemorrhage was found at autopsy. The cardiac repair was intact.

**Discussion**

It is now generally accepted that all infants with TGA should have an atrial septal defect created. In our hands the balloon technic has continued to be both a safe and effective procedure. Only rarely has a surgical excision of the septum been subsequently required. The atrial defect improves arterial oxygen content and reduces pulmonary venous hypertension. This latter factor may be important in limiting the development of pulmonary vascular disease in patients with TGA. One of our patients with simple TGA had PAH, but none has had increased pulmonary vascular resistance.

Since cerebral complications remain a serious hazard, particularly in patients with simple TGA even after BAS, an aggressive approach with early correction in this group is indicated. The low morbidity and mortality of total correction in small infants in this group lend support to this concept. Although a weight of 10 kg is almost ideal for total correction, we feel that the operation should be carried out in infants weighing less if there is evidence that the infant's rate of weight gain has decreased or his general condition is deteriorating. A CVA is clearly an indication for early correction. The results of physiologic correction in patients with simple TGA have been gratifying. The operative mortality is well below that of many other operations for major congenital heart lesions, although long-term results are as yet unknown.

Patients with TGA and a large VSD are likely to have early onset of PAH. Viles and associates have shown that many of these patients have severe pulmonary vascular changes after 6 months of age. BAS plus the aggressive use of early pulmonary artery banding has been advocated by our group

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and by Stark and associates. In patients with TGA and VSD, recatheterization is performed between the ages of 8 and 10 months. Patients with TGA, VSD, and persistent PAH then have PAB. Our data suggest that PAB will prevent the development of pulmonary vascular disease in patients with TGA and VSD. However, further experience is needed with recatheterization of these patients after complete correction, a difficult task after three or four previous studies have been performed. Removal of woven Teflon bands has not been a serious problem in contrast to that with the cotton fabric tape. In the latter cases band removal and time-consuming pulmonary artery reconstruction by patch graft angioplasty have been required.

Based on the experience reported above, we now believe that correction of the patient with TGA, VSD, and PAB should be performed after 4 years of age. Since CVA is uncommon in these patients, probably because of the improved mixing, there is not the urgency for correction as in patients with simple TGA. Closure of the VSD through the tricuspid valve is technically easier if the tricuspid annulus and the ventricular cavity are larger than is commonly seen in a 2-year-old. The transatrial approach to the VSD is far superior to an approach that necessitates a systemic ventriculotomy. The defects frequently have a muscular rim which holds sutures poorly. Two defects closed by mattress sutures backed with felt pledgets were observed to be partially open at autopsy. It is now our policy to close all VSD in these patients with a fabric patch to prevent tension on the rim and pulling out of sutures.

Our experience in patients with TGA and PS suggests that these patients can be managed much like infants with simple TGA. The added procedure of excising the subvalvular tissue at the time of total correction has not presented a serious problem. A systemic-pulmonary artery anastomosis may not be indicated. Two additional patients with TGA and PS developed severe congestive heart failure and died following ascending aorta-right pulmonary artery anastomosis although the anastomosis did not appear large. The one patient in our series who had total correction after an ascending aorta-right pulmonary artery anastomosis had a very small shunt which undoubtedly contributed little to pulmonary flow. We believe total correction rather than a palliative shunt should be performed.

Patients with TGA, VSD, and PS have proven to be the most difficult to manage. Each of the three patients in our series died in the postoperative period. These deaths cannot necessarily be attributed solely to the combination of lesions and their repair since two patients died with subarachnoid hemorrhage. Daicoff and his associates have reported excellent results using a method of repair similar to the one we have employed. On the other hand, a generally unfavorable experience with the direct repair of TGA, VSD, and PS led Rastelli and his associates to utilize an alternate method. They have closed the VSD with a contour patch that directs left ventricular output to the dextroposed aorta. The proximal end of the divided pulmonary artery is ligated. Right ventricular output is directed through a new pulmonary outflow tract and valve to the distal end of the pulmonary artery, using a homograft aorta and valve. Since this procedure requires insertion of the homograft of fixed size, waiting to the age of 5 years to perform total correction would seem wise. Should the patients have anoxic spells or an increasing hematocrit prior to that time, some type of systemic-pulmonary anastomosis can be performed in this group without excessive risk.

A flow diagram depicting our current plan of total management of patients with the major types of TGA is presented in Table 4.

Our initial impression that enlargement of the new physiologic left atrium with a diamond-shaped patch has led to a smoother postoperative course with fewer pulmonary complications has been confirmed by subsequent experience. Although three of ten patients who did not have the atrial enlargement had early evidence of pulmonary venous obstruction, none of the eight patients with
**Table 4**

*Management of Patients with Transposition of the Great Arteries*

<table>
<thead>
<tr>
<th>TGA</th>
<th>BAS</th>
<th>Isolated</th>
<th>VSD</th>
<th>PS</th>
<th>VSD, PS</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>1-2 yr (10 kg)</td>
<td>8 months</td>
<td>1-2 yr (10 kg)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Recatheterization, correction</td>
<td>Recatheterization</td>
<td>Recatheterization, correction</td>
<td>Systemic-pulmonary anastomosis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>8-10 months</td>
<td>With PAH</td>
<td>Without PAH</td>
<td>4-5 yr</td>
</tr>
<tr>
<td></td>
<td></td>
<td>4 yr (20 kg)</td>
<td>PAB</td>
<td>(15-20 kg)</td>
<td>Recatheterization, correction with homograft</td>
</tr>
</tbody>
</table>

*See text for abbreviations.*
simple TGA who had left atrial enlargement has had this complication.

References
Physiologic Correction of Transposition of the Great Arteries: Indications for and Results of Operation in 32 Patients

JOHN A. WALDHAUSEN, WILLIAM S. PIERCE, C. DICK PARK, WILLIAM J. RASHKIND and SIDNEY FRIEDMAN

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