Anatomic correction of simple transposition of the great arteries in 50 neonates

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ABSTRACT  From April 1984 to January 1986, anatomic surgical correction was performed in 50 newborn (2- to 23-day-old, mean 8 ± 5 [SD]) infants with simple transposition of the great arteries. Before surgery, balloon atrial septostomy was performed in all patients, prostaglandin E1 was infused in 42, and left ventricular shape on a two-dimensional echocardiogram was considered “satisfactory” in 48. Surgery was performed in patients on cardiopulmonary bypass without cardiac arrest; the pulmonary artery was reconstructed by end-to-end anastomosis according to Lecompte’s maneuver with a pericardial patch. In all but one patient coronary artery transfer was possible regardless of the distribution of these vessels. There were eight early deaths (16%), but only four (10%) of the last 41 patients treated died. There was one late death (2%) due to a secondary myocardial infarction caused by compression of the left coronary artery. Reoperation was successfully performed in two patients for supravalvar pulmonary artery stenosis. The only late medical complication was a transient episode of myocardial ischemia 6 months after surgery. The 41 late survivors were in excellent condition, were in sinus rhythm, and had a normal left ventricle 1 to 22 months after surgery (mean 7.2 ± 5.4 [SD]). Aortic growth was normal; pulmonary artery supravalvar stenosis occurred in six patients (mild in four). We conclude that anatomic correction can be applied successfully in the first few days of life in newborns with simple transposition of the great arteries, regardless of coronary distribution.


ANATOMIC CORRECTION of transposition of the great arteries (TGA) is theoretically a better therapy than atrial repair, since it does not introduce any additional intracardiac anomaly and it restores the left ventricle to its natural systemic function. But anatomic correction is a delicate operation requiring the transfer of the coronary arteries and reconstruction of the pulmonary artery. The long-term fate of these vascular structures remains unknown. Since the first successes were reported in 1975,1,2 anatomic correction appeared for the majority of pediatric cardiology centers as the method of choice in managing TGA with ventricular septal defect (VSD).3,4 The use of anatomic correction for simple TGA is more questionable for two main reasons: (1) atrial repair5 of simple TGA provides excellent immediate and satisfactory midterm results, and (2) anatomic repair carries an important risk of failure because of the natural underdevelopment of the left ventricle, which soon after birth becomes a low-pressure ventricle. Therefore, anatomic correction has to be performed either after preliminary preparation of the left ventricle, or in the newborn.

A two-stage procedure for treatment of TGA was introduced and successfully developed by Yacoub et al.,6,7 but in our hands this method failed, mainly because of high mortality and morbidity associated with the first stage of pulmonary arterial banding.8,9 Furthermore, this preliminary stage may alter left ventricular function,10 and may dilate the future proximal aorta,11,12 inducing aortic insufficiency.3,11 Encouraged by the successful report by Hougen et al.13 in Boston, in April 1984 we decided to attempt to treat simple TGA by anatomic correction in the neonatal period. Well-informed parental consent was obtained in each case and the protocol was applied only to a selected group of patients who were less than 2 weeks old and had a suitable left ventricle. We report here our results with this approach.
Patients and methods

From April 1984 to January 1986, 50 newborn (2- to 23-days-old, mean ± SD = 8 ± 5) (mean values are ± SD throughout) infants were referred to the same surgeon for anatomic repair of TGA with intact (or almost intact) ventricular septum.

Preoperative data. All patients underwent catheterization with balloon atrial septostomy, 47 at less than 5 days of age and three at between 10 and 13 days of age. Left and right ventricular angiograms showed an intact ventricular septum in all but three patients with trivial VSDs in the trabecular septum. The left/right ventricular systolic pressure ratio was greater than 0.6 in all patients (mean 0.78 ± 0.07). Prostaglandin (PG) E₂ was infused in 36 patients at the time of the investigation and was started in 11 others later to maintain high pulmonary arterial and left ventricular pressures. Only three patients who were treated at the beginning of the series did not receive PGE₂; in these patients operation was performed before the age of 4 days. In six patients, PGE₂ was discontinued before the operation because it was not well tolerated. Our goal was to operate in infants about 1 week old and definitely before 2 weeks of age, but in four instances surgery could not be undertaken before 15 days, and the longest delay was 23 days. Thus, delay between atrial septostomy and anatomic correction was less than 10 days in 41 patients, was between 10 and 15 days in eight, and was more than 15 days in one.

All patients were carefully evaluated before operation by daily two-dimensional echocardiography to assess left ventricular size, wall thickness, contraction, and shape. The end-systolic left ventricular shape was assessed in the subxiphoid transverse view (figure 1) and was considered a rough but reliable estimation of the left ventricular systolic pressure. Thus, left ventricular geometry was considered to be favorable (type 1) when the lateral/anteroposterior diameter ratio was equal to or less than 2/1, acceptable (type 2) when the ratio was between 2/1 and 3/1, and unfavorable (type 3) when the ratio was equal to or more than 3/1. Table 1 summarizes the preoperative echocardiographic data and shows that left ventricular geometry worsened with increasing age and improved with the infusion of PGE₂.

In five patients, anatomic correction was performed despite an unfavorable age or left ventricular shape: two had a favorable (type 1) left ventricular shape after undergoing a late septostomy at 13 and 16 days of age and were operated on again at 16 and 19 days, one had massive tricuspid insufficiency contraindicating atrial repair and was operated on at 23 days of age even though the left ventricular shape was unfavorable, and in the last two, the parents asked specifically for anatomic correction despite the age and the left ventricular shape.

During the same period, eight other patients were treated by atrial repair because of unfavorable left ventricular geometry and/or age (seven patients) or frontal position of the great arteries (one patient).

Surgical data. Surgery was performed in patients on cardio-pulmonary bypass under hypothermia (19° to 23°C), but without cardiac arrest. Mean aortic cross-clamping time was 67 min. The transfer of the coronary ostia to the posterior vessel was accomplished according to the technique described by Yacoub and Radley Smith. The coronary artery distribution was type A (normal) in 32, type B or C (single artery arising from a posterior sinus, or two vessels very close to each other and to the posterior commissure) in six, type D (two vessels with a circumflex artery arising from the right coronary artery posteriorly) in nine, and type E (two vessels with a main descending artery arising from the right coronary artery) in three. The reconstruction of the pulmonary artery was performed in all by end-to-end anastomosis as described by Lecompte et al. with the use of pericardium to fill the gaps in the posterior wall of the original aorta caused by the excision of the coronary ostia. Two patches were used to close each gap separately in the first 10 patients, and one large rectangular patch was used in the last 40. Tanned pericardium was used in the first 25 patients; native autologous pericardium was used in the last 25.

Postoperative methods. Surgical survivors had follow-up visits at 1, 2, 3, and 6 months after surgery, and every 6 months thereafter. Clinical, chest x-ray, and standard electrocardiographic examinations were performed at each visit. Complete M mode and two-dimensional echocardiographic examinations were also done to assess left and right ventricular function, annular size, and supravalvular anastomotic areas. The echocardiographic measurements of surface area were compared with

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**TABLE 1**

| Immediate preoperative echocardiographic data from 50 patients with simple TGA |
|------------------|------------------|------------------|
|                  | LV diameter (mm) | LVFW thickness (mm) | Septal geometry³ |
|                  | n    | ED±SD | ES±SD | n    | ED±SD | ES±SD | Type 1 | Type 2 | Type 3 |
| Age (days)       |      |       |       |      |       |       |        |        |        |
| 2-9              | 36   | 14.2±3.1 | 6.1±1.1 | 3.3±0.6 | 6.1±1.1 | 22   | 14   | 0     |
| 10-23            | 14   | 16.7±2.3 | 6.5±1.1 | 3.1±0.4 | 6.5±1.1 | 3    | 9    | 2     |
| With PGE₁ infusion | 42  | 16.2±2.1 | 6.4±1.3 | 3.2±0.6 | 6.3±1.0 | 25   | 17   | 0     |
| Without PGE₁ infusion | 8   | 13.9±2.2 | 5.9±1.6 | 3.2±0.5 | 6.4±1.1 | 0    | 6    | 2     |

All values are means ± 1 SD.

ED = end-diastole; ES = end-systole; LV = left ventricular; LVFW = left ventricular free wall.

³See text for definition of types.
Length of follow-up ranged from 1 to 22 months for the 42 early survivors, with a mean of $7.25 \pm 5.4$ months.

There was one late death from a severe myocardial infarction 6 weeks after an uneventful operation in a patient with a type A coronary distribution. At autopsy, there was an extrinsic compression of the left coronary artery by fibrous tissue around the tanned pericardial patch used to reconstruct the pulmonary artery. Another child suffered a localized myocardial infarction in the distribution of the right coronary artery 6 months postoperatively. He completely recovered in 3 weeks and is doing well 6 months later with a normal electrocardiogram, normal echocardiogram, and two apparently normal coronary arteries on the angiogram. Two patients were reoperated upon 12 and 14 months after their anatomic correction because of a supravalvular pulmonary arterial stenosis due to retraction of the tanned pericardial patch, a small pulmonary arterial anulus, and dysplastic pulmonary arterial valves: both are in excellent condition 3 and 6 months later.

No complications were noted in the 38 other patients and all 41 survivors are free of symptoms, with normal growth and psychomotor development. They have no heart failure and normal heart size on the chest x-ray. Six have a significant systolic murmur ($\geq 3/6$) due to pulmonary arterial stenosis, and one has a minute diastolic murmur due to aortic regurgitation (table 2). The electrocardiogram shows ischemic signs in only two early postoperative patients who have undergone a very short follow-up (1 month). All patients are in sinus rhythm and 24 hr recordings are normal. Right ventricular hypertrophy decreased rapidly after surgery, with left ventricular forces becoming predominant by 1 month, and a normal electrocardiogram was recorded by 3 months in all but the six patients who developed pulmonary arterial stenosis (table 3).

On the echocardiogram, the left heart was always found to be normal; it was never dilated or hypokinetic (table 2). In six patients, the left ventricle was hyper-
TABLE 3
Postoperative results — right heart

<table>
<thead>
<tr>
<th>Length of follow-up (months)</th>
<th>n</th>
<th>SM≥grade III/IV</th>
<th>RVH (ECG)</th>
<th>ED diameters (RV/LV≥0.5)</th>
<th>Septal geometry&lt;sup&gt;a&lt;/sup&gt;</th>
<th>Type 1</th>
<th>Type 2'</th>
<th>Type 3'</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;3</td>
<td>11</td>
<td>0</td>
<td>5</td>
<td>0</td>
<td></td>
<td>0</td>
<td>2</td>
<td>9</td>
</tr>
<tr>
<td>≥3</td>
<td>30</td>
<td>6</td>
<td>6</td>
<td>5</td>
<td></td>
<td>2</td>
<td>4</td>
<td>21</td>
</tr>
</tbody>
</table>

ED = end-diastolic; LV = left ventricle; RV = right ventricle; RVH = right ventricular hypertrophy; SM = systolic murmur; ECG = electrocardiography.

<sup>a</sup>See text for definition of types. Not available in three patients.

kinetic, with relative hypertrophy and low end-systolic wall stress. The aortic root was never significantly dilated and we never observed supravalvular stenosis (table 3). The right heart was abnormal in six patients (table 3), with mild dilatation of the right ventricle in five instances and abnormal end-systolic left ventricular geometry in all, suggesting high right ventricular systolic pressures. The pulmonary arterial ring was small in two patients and supravalvular pulmonary arterial stenosis was present in all six. On the other hand, we did not find any case of stenosis of the origin of a pulmonary branch among the 33 patients in whom it could be examined.

Right heart catheterization confirmed the existence of pulmonary arterial stenosis in the six patients with loud systolic murmur, persistent right ventricular hypertrophy on the electrocardiogram, and abnormal echographic left ventricular shape. This lesion was severe in one child (right ventricular pressure 110 mm Hg), moderate in another (70 mm Hg), and mild in the remaining four (40 to 60 mm Hg). The site of the stenosis was supravalvular in all patients, with additional valvular stenosis in two, but none had pulmonary arterial branch stenosis. The angiographic levophase was normal in the 10 catheterized patients (figures 2 to 4).

Discussion

This study confirms that anatomic correction of simple TGA can be performed successfully in the neonate. Distribution of the coronary arteries and spatial relations of the great vessels were only occasionally of importance: in only one case did the correction appear impossible due to a single coronary ostium located in the posterior commissure; this situation is very difficult to identify preoperatively. When compared with the two-stage procedure of repair, not only was the early mortality lower for neonatal anatomic correction, but the quality of the repair also seemed to be considerably improved because the deleterious effects of pulmonary arterial banding on the future ascending aorta<sup>3,12</sup> and eventually on the left ventricular function<sup>10</sup> was avoided. Aortic leak, which is so common after a two-stage procedure in patients with complex<sup>3</sup> or simple TGA<sup>12</sup> together with dilatation of the proximal aorta, occurred only once in our present series. In this patient, regurgitation was present from the first postoperative day and appeared to be secondary to a technical error and not to aortic root dilatation. Although immediate postoperative ischemic problems were common, they were always
FIGURE 3. Supravalvular pulmonary arterial stenosis. Right ventricular angiography in the four-chamber view (A) and in the lateral view (B). Tight stenosis of the pulmonary arterial trunk from the annulus up to the bifurcation 11 months after surgery. Right ventricular systolic pressure was 100 mm Hg, an indication for subsequent surgery.

transitory and left ventricular function was always normal.

Our midterm follow-up data lead us to believe that two main problems remain matters of concern. The first is the occurrence of late coronary failure, which led to the only late death in our experience and resulted from extrinsic compression of the left coronary artery. Such complications have not been reported to our knowledge in the late follow-up of anatomic correction of TGA, but no selective coronary angiograms have been performed in previous studies. Thus, the long-term future of these transferred vessels is still unknown, even though there are experimental data suggesting that normal growth is likely to occur.19

The second problem concerns the fate of the new pulmonary artery. The pulmonary artery was repaired by one or two patches after the coronary buttons were removed, leaving little normal tissue with the ability to grow between the new aortic posterior sinuses. Although we have had only two cases of moderate-to-severe pulmonary stenosis compared with 35 of 41 patients free of this complication, our follow-up was too short to assess the quality of the pulmonary arterial growth and the real incidence of this complication in the future. It is possible that pulmonary arterial stenosis was due to retraction of the tanned pericardium since we have not observed any case of stenosis among the last 25 patients who were treated with autologous pericardium. But since the follow-up period of this last subset of patients is the shortest, we are not able to draw conclusions. We observed no pulmonary arterial branch stenosis after decrossing of the pulmonary artery, so we believe that this is a better approach to reconstruction of the pulmonary artery than the use of conduits20 that will have to be changed at least once during growth.

Finally, the selection of patients, the optimal age, and the best preparation for neonatal anatomic correction should be considered. We selected our patients according to age and left ventricular shape, but we are aware that these criteria are arbitrary. It seems that left ventricular thickness and mass remain in the range of normal values for at least 1 month after birth in infants with simple TGA,21,22 and that a low left ventricular systolic pressure, as suggested by unfavorable left ventricular shape, only means that pulmonary arterial
pressure has decreased with the physiologic decrease of pulmonary arterial resistances, and does not necessarily indicate that the left ventricle has lost its ability to develop high pressure. However, we cannot disregard the fact that the two patients who died without problems of the coronary arteries or myocardial preservation were among the oldest (15 and 23 days) and had unfavorable left ventricular shapes (types 2 and 3). On the other hand, the two other patients operated on at 16 and 19 days of age who had favorable left ventricular geometry, and the 13-day-old baby with an unfavorable left ventricular geometry, had remarkably simple postoperative courses and no left ventricular failure. It is thus possible that fulfillment of both criteria is not mandatory and that the very young patient can be a candidate for anatomic correction regardless of the left ventricular shape or pressure, and that the patient with favorable left ventricular shape or pressure can be a candidate for this operation regardless of his age.

Optimal age for operation appears to be around the age of 1 week, because it leaves time to assess extra-cardiac problems (neonatal infection for instance) and for major cardiovascular and pulmonary changes to take place, while it does not leave time enough for the left ventricle to lose its ability to develop high pressure. We also believe that the patient should undergo a balloon atrioseptostomy that decreases pulmonary edema and improves systemic oxygenation. In addition, our results indicate (table 1) that the infusion of PGE, started after septostomy maintains left ventricular preparation by increasing flow and pressure in the pulmonary bed without pulmonary edema.

References


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