Anatomic Correction of Transposition of the Great Arteries: Pre- and Postoperative Cardiac Catheterization, with Angiocardiography in Five Patients

ROBERT M. FREEDOM, M.D., J. A. G. CULHAM, M.D., PETER M. OLLEY, M.D., RICHARD D. ROWE, M.D., WILLIAM G. WILLIAMS, M.D., AND GEORGE A. TRUSLER, M.D.

SUMMARY Six of eight patients survived anatomic correction of transposition of the great arteries and repair of associated cardiovascular anomalies at the Hospital for Sick Children, Toronto, Canada. Two of the six survivors also had tricuspid atresia, and continuity between the right atrium and the subaortic outlet chamber in these patients was provided by a valved external conduit, in addition to the arterial switch and coronary artery reimplantation. Five of the six patients have undergone complete postoperative clinical, hemodynamic and angiocardiographic investigation and form the basis of this communication. Our present indications for anatomic repair are discussed.

CONSIDERABLE EXCITEMENT was generated by the report of Jatene et al. in 1975 of successful anatomic correction of transposition of the great arteries. Subsequently, techniques for anatomic correction were independently developed, applied and published by several groups. Yet, with the exception of the large experience of Yacoub et al. other reports have largely been anecdotal. We report the pre- and postoperative catheterization findings in five patients who survived anatomic correction of transposition of the great arteries at the Hospital for Sick Children in Toronto and discuss our present indications for this approach.

Methods

Patients

Between August 1978 and April 15, 1980, eight patients, ages 9 days to 14 years, 11 months underwent an arterial switch with coronary artery reimplantation and correction of associated anomalies at the Hospital for Sick Children. Six of the eight patients survived, and five of the six have undergone complete postoperative clinical, hemodynamic and angiocardiographic assessment and form the basis of this report. One recently operated patient, a 5-month-old infant with complete transposition of the great arteries and a large ventricular septal defect, survived and had an excellent postoperative course, but has not yet undergone complete postoperative catheter assessment.

Two patients, a 9-day-old baby who had complete transposition of the great arteries and a huge patent ductus arteriosus and a 3-year-old who had a Taussig-Bing double-outlet right ventricle and an infundibular ventricular septal defect, did not survive operation. The anatomic diagnoses in the five survivors who had complete postoperative assessment are given in table 1.

The timing of surgery was dictated by hypoxia, congestive heart failure and pulmonary hypertension in five of the eight operated patients, two of whom (patients 2 and 3) had complete postoperative assessment. The indications for anatomic correction were exertional dyspnea, mild pulmonary artery hypertension, left ventricular dysfunction and potential subaortic stenosis in patient 4, and hypoxia and increasing polycythemia in patient 1. At catheterization 3 years after banding of the pulmonary artery, patient 5 was found to have subaortic stenosis secondary to restriction at the outlet foramen. The success of anatomic correction in a similar patient (no. 4) persuaded us to take this approach.

Results

Preoperative Hemodynamic Investigation (table 1)

The systemic arterial oxygen saturation ranged from 59–86% at catheterization immediately before surgery. The morphologic right and left ventricles were both at systemic pressures. Pulmonary artery pressure equalled systemic pressure in the three patients without a previously banded pulmonary artery. Of the three patients with a banded pulmonary artery, the pulmonary artery pressure distal to the band was normal in patients 1 and 5 and mildly elevated in patient 4, who had a torrential pulmonary blood flow.

Preoperative Angiocardiographic Findings

In patient 2, the only patient with complete transposition and a large patent ductus arteriosus, right

From the Division of Cardiology, Departments of Pediatrics, Pathology and Radiology, and the Division of Cardiovascular Surgery, Department of Surgery, The Hospital for Sick Children, and the Departments of Pathology, Pediatrics and Surgery, The University of Toronto Faculty of Medicine, Toronto, Ontario, Canada.

Address for correspondence: Robert M. Freedom, M.D., The Hospital for Sick Children, 555 University Avenue, Toronto, Ontario, Canada M5G IX8.

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ventricular function was depressed and the upper interventricular septum bulged paradoxically into the left ventricle during right ventricular systole. The right ventricular ejection fraction in this patient was 40%, and the left ventriculogram showed preferential flow into the right pulmonary artery. A discrepancy in the pulmonary root and aortic root was observed in three patients. Finally, no significant difference in root dimension was observed in the patient with Taussig-Bing double-outlet right ventricle and previously banded pulmonary artery, although the left ventricular semilunar valve (pulmonary valve) had perhaps a slightly larger circumference.

Operative Technique

The operative technique for an arterial switch repair of transposition has been reported and continues to evolve. The technique for repair with tricuspid atresia is the subject of a separate report. In general, we have maintained sufficient length of the distal aorta and proximal pulmonary artery to allow direct anastomosis of these two structures without undue tension or intervening grafts. To overcome most of the discrepancy between aortic and pulmonary artery diameters, the aortic incision can be oblique. In one patient, the oblique aortotomy incision was extended under the right coronary artery, thereby keeping the origin of the right coronary in continuity with the aorta and simplifying subsequent reimplantation. The coronary arteries are sectioned after the aortic incision has been made and the heart protected by a combination of hypothermia and cardioplegia. The proximal 5-10 mm of the coronary arteries are mobilized, and branches can be seen from inside the coronaries both directly and with probes. This is usually sufficient to allow the posterior rotation of the coronary ostia to the adjacent pulmonary artery. The site of reimplantation for the coronary arteries should be away from the pulmonary annulus because a low-lying incision can weaken the support structure of valve, as in patient 4. The site of reimplantation should be somewhat lateral on the right side to avoid compression of the coronary artery by the overlying pulmonary artery reconstruction, which may have contributed to coronary stenosis seen in patient 2. Ventricular septal defects, particularly in the presence of double-outlet right ventricle, can be closed conveniently through the pulmonary valve, as in both patients with a double-outlet right ventricle. Two children with ventricular septal defects underwent closure through the right ventricle and one through the right atrium. In the two patients with tricuspid atresia, the ventricular septal defects were closed through a ventriculotomy in the outlet chamber.

Reconstruction of the pulmonary artery required a tubular graft in all but one patient. The graft must connect the anatomic aortic valve to the pulmonary artery bifurcation and therefore must pass around one-half of the circumference of the ascending aorta, a distance of 2-5 cm. Even in infants, the graft can be large enough to accommodate subsequent growth. The conduit sizes varied from 15-30 mm in diameter. In three of the six survivors, the graft passed to the right of the reconstructed ascending aorta and in three, the graft passed to the left.

Table 1. Hemodynamic Data

<table>
<thead>
<tr>
<th>Pt</th>
<th>Procedure</th>
<th>Oxygen saturation data (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>SVC</td>
</tr>
<tr>
<td>1</td>
<td>A. Taussig-Bing double-outlet right ventricle, banded pulmonary artery, age 5 weeks</td>
<td>59</td>
</tr>
<tr>
<td></td>
<td>B. Post pulmonary artery banding &amp; SE, age 2 yrs, 10 mos</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>C. Post Jatene, 4 mos &amp; VSD closure, age 3 yrs, 6 mos</td>
<td>74</td>
</tr>
<tr>
<td>2</td>
<td>A. TGA &amp; PDA, age 10 days</td>
<td>60</td>
</tr>
<tr>
<td></td>
<td>B. 1 year after repair, age 20 mos</td>
<td>70</td>
</tr>
<tr>
<td>3</td>
<td>A. TGA &amp; VSD, age 8 mos</td>
<td>59</td>
</tr>
<tr>
<td></td>
<td>B. Age 9.5 mos</td>
<td>68</td>
</tr>
<tr>
<td>4</td>
<td>A. TGA, TAT, banded pulmonary artery, age 29 mos</td>
<td>56</td>
</tr>
<tr>
<td></td>
<td>B. Age 8 yrs, 7 mos</td>
<td>53</td>
</tr>
<tr>
<td></td>
<td>C. Age 14 yrs, 4 mos</td>
<td>64</td>
</tr>
<tr>
<td></td>
<td>D. 1 mo postop, age 15 yrs</td>
<td>65</td>
</tr>
<tr>
<td>5</td>
<td>A. TGA, TAT, banded pulmonary artery, restrictive VSD, age 2 mos</td>
<td>44</td>
</tr>
<tr>
<td></td>
<td>B. Age 8 yrs, 9 mos</td>
<td>62</td>
</tr>
<tr>
<td></td>
<td>C. Age 9 yrs, 4 mos, 2 mos after repair</td>
<td>58</td>
</tr>
</tbody>
</table>

Abbreviations: DORV = double-outlet right ventricle; SE = Sterling-Edwards procedure; PA = pulmonary artery; TAT = tricuspid atresia; VSD = ventricular septal defect; TGA = transposition of the great arteries; PDA = patent ductus arteriosus; SVC = superior vena cava; IVC = inferior vena cava; RA = right atrium; RV = right ventricle; LA = left atrium; LV = left ventricle; MPA = main pulmonary artery; AO = aorta; NE = nonexistent; NM = not measured.
Deaths

One patient with a Taussig-Bing double-outlet right ventricle and pulmonary artery hypertension died intraoperatively. This 3-year-old boy was found at necropsy to have a good anatomic repair, but the cause of death was related to pulmonary vascular arteriopathy and an unrecognized mild thoracic coarctation of aorta that was unmasked and unrecognized after intraoperative ligation of a large patent ductus arteriosus. The youngest patient in this series died at 9 days of age; at necropsy, his left ventricle was 4.0 mm thick and the ostium of his left coronary artery was quite narrow.

Postoperative Clinical Assessment

All five patients who underwent complete postoperative assessment looked well and were clinically acyanotic. All five had systolic ejection murmurs thought to emanate from the right ventricular outflow tract. One patient had a soft pansystolic murmur at the lower left sternal border that was attributed to a residual ventricular septal defect. High-frequency early diastolic murmurs of aortic incompetence were heard in three patients. The chest radiograms showed a decreased cardiothoracic ratio in all patients. The pulmonary markings approached normal.

Postoperative ECG

All five patients remained in normal sinus rhythm postoperatively, although one patient had first-degree atrioventricular block (PR interval 0.22 second). Two patients had an rSR' pattern in V1 and two patients had complete right bundle branch block. The frontal QRS axes in the patients with transposition and double-outlet right ventricle remained inferior or rightward. No patient developed left-axis deviation and complete right bundle branch block.

Postoperative Hemodynamic Assessment

The arterial oxygen saturation ranged from 92–99%. A residual right-to-left shunt was excluded in all patients on the basis of forward dye curves. Patients 1 and 3 had trivial left-to-right shunts at the ventricular level detected only by reverse dye curve and by selective left ventricular cineangiography (fig. 1D). A pressure gradient of more than 10 mm Hg across the right ventricular outflow tract was recorded only in patient 1 and none of the patients had a pressure gradient of more than 10 mm Hg across the left ventricular outflow tract. Stenosis of the main pulmonary artery at the anastomotic site between the Dacron graft and distal pulmonary artery was identified in patient 1, who had a gradient of 47 mm Hg across the right ventricular outflow tract. The aortic pulse pressure was normal in the four patients, despite angiographic evidence of mild aortic incompetence.

Postoperative Angiographic Assessment

Moderately severe right ventricular outflow obstruction was identified only in patient 1, in whom the morphologic right ventricle was hypercontractile (figs. 1A–C); the lateral right ventriculogram shows the point of obstruction (fig. 1C). A long Dacron conduit was necessary to provide continuity between the morphologic right ventricle and the pulmonary artery in patient 3 (fig. 2A). The discrepancy between the right ventricular semilunar valve and pulmonary artery is shown in figure 3A.

Mild aortic incompetence was identified in patients 2–5. In patients 3 and 4 there was a significant

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**Table 1. (Continued)**

<table>
<thead>
<tr>
<th>Oxygen saturation data (%)</th>
<th>Pressure data (mm Hg)</th>
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<tbody>
<tr>
<td>LA</td>
<td>LV</td>
</tr>
<tr>
<td>----</td>
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</tr>
<tr>
<td>95</td>
<td>92</td>
</tr>
<tr>
<td>90</td>
<td>87</td>
</tr>
<tr>
<td>NE</td>
<td>92</td>
</tr>
<tr>
<td>95</td>
<td>91</td>
</tr>
<tr>
<td>NE</td>
<td>99</td>
</tr>
<tr>
<td>96</td>
<td>95</td>
</tr>
<tr>
<td>91</td>
<td>89</td>
</tr>
<tr>
<td>NM</td>
<td>81</td>
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<tr>
<td>84</td>
<td>86</td>
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<td>88</td>
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<td>94</td>
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<tr>
<td>100</td>
<td>98</td>
</tr>
<tr>
<td>65</td>
<td>83</td>
</tr>
<tr>
<td>NE</td>
<td>96</td>
</tr>
</tbody>
</table>
difference in size between the left ventricular semilunar valve and the surgically anastomosed ascending aorta (figs. 1E and F, 2B–D, 3B and C, 4A–C), although there was no hemodynamic evidence of obstruction in the supravalvular aortic area. In one patient, although the roots were comparable, aortic incompetence was still found postoperatively (figs. 4A and B).

The ostium of the right coronary artery was narrowed in patient 2 (fig. 4C), and in patient 4, the ostium of the left coronary artery was narrowed.

Discussion

In 1964, a new era for the patient with complete transposition of the great arteries was opened with Mustard’s atrial partitioning operation. However, the results of postoperative hemodynamic, angiographic and electrophysiologic evaluation of survivors of this inflow “correction” or atrial partitioning, began to raise questions about the efficacy of this operation. Although initially some technical concerns were raised with regard to the caval or pul-
monary venous obstruction and the material used in partitioning the atria (pericardium or Dacron),\textsuperscript{20} longitudinal follow-up studies of large numbers of these patients have reported the occurrence, incidence and natural history of supraventricular dysrhythmias and their electrophysiologic basis\textsuperscript{21} and right ventricular dysfunction.\textsuperscript{22-26} Attempts to lessen the incidence of supraventricular dysrhythmias by modifying the shape of the atrial baffles have been described, and a return to Senning’s procedure has also been advocated.\textsuperscript{22-25} Hagler et al.,\textsuperscript{26} reviewing their patients who survived atrial partitioning, observed that significant depression of right ventricular function was likely to result in substantially diminished cardiac reserve and reduced survival, and concluded that these observations supported use of the left ventricle as the systemic ventricle in correction of complete transposition of the great arteries.

Yet, more than 25 years have passed since Mustard’s publication of attempted and unsuccessful anatomic correction of transposition before this approach was used.\textsuperscript{96} Anatomic correction has both advantages and disadvantages (table 2).\textsuperscript{3, 97} The potential for coronary ostial stenosis, supravalvular aortic and pulmonary stenosis and aortic incompetence is a major concern with anatomic correction and coronary artery reimplantation. Although we have not observed acquired supravalvular aortic stenosis in our small series, this complication would be expected when anastomosing relatively small vessels of different circumferences. Both coronary ostial stenosis and aortic incompetence have occurred in our patients and have been reported by others. Longer follow-up with complete hemodynamic and angiographic evaluation, and selective coronary arteriography, when necessary, in a large cohort of patients treated by
anatomic correction and coronary artery reimplantation may yield a “Pandora’s box” of complications. Yet, the complications anticipated from the anatomic correction are vastly different from those resulting from an “inflow” approach for correction (table 3).5, 37-40

We have not taken the position advocated by Radley-Smith and Yacoub that anatomic correction
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with coronary artery reimplantation is the surgical procedure of choice for the patient with complete transposition and intact ventricular septum with "normal" left ventricular and pulmonary artery pres-

ures.²⁴ Among patients with complete transposition of the great arteries and intact ventricular septum and normal left-sided pressures, an inflow type of operation (Mustard or Senning) can be performed with an operative mortality of 2%, considerably less than that reported in the two-stage procedure required for anatomic correction.²⁰ The 5-year actuarial survival for these patients in our institution is 91%. Second, we are well aware of the atrial rhythm disturbances intrinsic to any type of inflow correction, but further modifications of operative technique could lessen this complication, which in any case is often of minor impact. Further, it is unclear whether all of the cardiac arrhythmias documented after the Mustard operation are truly acquired.⁴¹

TABLE 2. Potential Complications of AnatomicCorrection and Coronary Artery Reimplantation for Transposition of theGreat Arteries

<table>
<thead>
<tr>
<th>Complication</th>
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<tbody>
<tr>
<td>Acquired supravalvar aortic stenosis.</td>
</tr>
<tr>
<td>Coronary ostial stenosis.</td>
</tr>
<tr>
<td>Stenosis of Dacron conduit between right ventricular semilunar valve and distal pulmonary artery.</td>
</tr>
<tr>
<td>Aortic incompetence.</td>
</tr>
</tbody>
</table>

FIGURE 3. Postoperative angiograms in patient 4, who had tricuspid atresia, transposition of the great arteries, a previously banded pulmonary artery and impaired left ventricular function, and underwent arterial switch, coronary artery reimplantation, patch closure of atrial and ventricular septal defects, and reconstitution of right atrial-right ventricular continuity with a valved external conduit. (A) Lateral angiogram performed in reconstituted right ventricle ("RV") shows connection with the much enlarged pulmonary artery (PA). There is a striking discrepancy (white arrow) between the semilunar valve of the reconstituted right ventricle and the pulmonary artery. (B, C) Anteroposterior and lateral retrograde aortograms show a discrepancy in dimension between the left ventricular semilunar valve (LVS V) and the reconstituted aorta (AO). The aortogram shows mild aortic incompetence (curved arrow).
Figure 4. Postoperative angiocardiograms in patient 2, who had transposition of the great arteries, impaired right ventricular function and a patent ductus arteriosus. (A, B) Lateral and left anterior oblique retrograde aortograms show an eccentric aortic valve with aortic regurgitation into the morphologic left ventricle. (C) Selective right coronary arteriogram shows a narrowed proximal coronary artery. AO = aorta.

Table 3. Potential Complications of Atrial Partitioning for Transposition of the Great Arteries*

<table>
<thead>
<tr>
<th>Complication</th>
<th>Incidence (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena caval obstruction</td>
<td>4</td>
</tr>
<tr>
<td>Inferior vena caval obstruction</td>
<td>0.25</td>
</tr>
<tr>
<td>Pulmonary venous obstruction</td>
<td>3</td>
</tr>
<tr>
<td>Partial or complete loss of reservoir and contractile function of the atria</td>
<td>Unknown</td>
</tr>
<tr>
<td>Right ventricular dysfunction</td>
<td>≥ 30</td>
</tr>
<tr>
<td>Dynamic subvalvular pulmonary obstruction</td>
<td>&lt; 5</td>
</tr>
<tr>
<td>Abnormal distribution of pulmonary blood flow**</td>
<td>5</td>
</tr>
<tr>
<td>Atrial dysrhythmia</td>
<td>~ 30</td>
</tr>
</tbody>
</table>

*Based on clinical examination and postoperative cardiac catheterization of 205 patients with complete transposition and intact ventricular septum.
The etiology of right ventricular dysfunction in the patient with complete transposition is unclear.\textsuperscript{22-27} Although Becu and Gallo\textsuperscript{49} suggested that the right ventricular myocardial architecture in hearts obtained from patients with complete transposition is different from normal, it is difficult to extrapolate these differences in form to progressive alterations in function. Thus, it is uncertain whether the matrix for right ventricular dysfunction is congenital or acquired or both. The relative contributions of hypoxemia (before atrial partitioning), the loss of atrial function (after atrial partitioning), and the lifetime consequence of increased afterload (the systemic resistance before and after atrial partitioning) to a disordered right ventricular myocardium are unknown.\textsuperscript{9} Clearly, some patients have right ventricular dysfunction before atrial partitioning, and in others it develops and may progress after atrial partitioning.\textsuperscript{22-27} However, not all patients with complete transposition have obvious right ventricular dysfunction after atrial partitioning. Whether it will develop in such patients is conjectural, and longitudinal studies are needed to define the timing and incidence of this occurrence. We agree, however, that the severely impaired right ventricle (before any form of "corrective" surgery) would benefit from afterload reduction. Finally, inadequate myocardial protection at the time of operative repair may contribute to right ventricular dysfunction. Although myocardial preservation techniques are improving, some damage may be an inherent risk and may affect either ventricle.

Because our experience with the arterial switch and coronary artery reimplantation is small, we believe it unwise to formulate rigid guidelines for patient selection. Indications for this approach include complete transposition of the great arteries and severely depressed right ventricular function, and complete transposition of the great arteries and right ventricular hypoplasia (with or without a ventricular septal defect). In the patient without a ventricular septal defect, the left ventricle must be prepared. Preliminary data from several centers suggest that the patient with transposition of the great arteries and a large ventricular septal defect or the patient with a Taussig-Bing double-outlet right ventricle is at particular risk to develop right ventricular dysfunction.\textsuperscript{28-37, 48} However, such patients should be considered as candidates for the arterial switch and coronary artery reimplantation. Finally, we consider patients with tricuspid atresia, transposition of the great arteries, banded pulmonary artery and well-developed infundibular chambers as candidates for the arterial switch, coronary artery reimplantation and establishment of right atrial-ventricular continuity,\textsuperscript{16} although other surgical approaches can be used in these patients.\textsuperscript{44, 48} Because such patients are at considerable risk to develop "subaortic stenosis" resulting from progressive narrowing of the outlet foramen,\textsuperscript{47-49} anatomic repair should effectively treat or prevent this complication, as in the 9-year-old girl who survived anatomic repair and a Fontan-like operation. The first patient treated this way in our institution (no. 4) had moderate impairment of left ventricular function before anatomic repair. Although no pressure gradient was recorded across the outlet foramen, the defect appeared considerably narrower than the aortic root and was potentially obstructive. Therefore, anatomic repair, combined with a Fontan-like operation, seemed reasonable.

The coronary artery anatomy of patients with complete transposition has received considerable attention during the past 20 years,\textsuperscript{50-52} and since the successful anatomic correction of transposition by Jatene et al.,\textsuperscript{1} this interest has increased.\textsuperscript{53} Although Hvass suggested that direct bilateral reimplantation of the coronary arteries seemed possible in only 30 of 100 necropsy specimens,\textsuperscript{53} Yacoub and Radley-Smith state that coronary artery anatomy does not preclude successful anatomic correction.\textsuperscript{54}

In our institution, an absolute contraindication to the arterial switch and coronary artery reimplantation is the presence of significant valvar or subvalvar pulmonary stenosis, although the unique approach described by Bex and his colleagues obviates this concern.\textsuperscript{55} Left ventricular hypoplasia or significant anomalies of the left atroventricular junction also preclude anatomic correction.

In conclusion, we are not rigid in our adherence to atrial partitioning. There is evidence that anatomic correction with coronary artery reimplantation has a role in the treatment of carefully selected patients with complete transposition of the great arteries. However, for most patients with complete transposition, i.e., those with an intact ventricular septum or a small ventricular septal defect, we will not abandon the atrial partitioning approach until we are persuaded that the arterial switch with coronary artery reimplantation performed in infancy offers such patients a more favorable postoperative course.

References

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Anatomic correction of transposition of the great arteries: pre- and postoperative cardiac catheterization, with angiocardiography in five patients.
R M Freedom, J A Culham, P M Olley, R D Rowe, W G Williams and G A Trusler

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