Intracardiac Repair of Cardiac Malformations with Atrioventricular Discordance

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SUMMARY Twenty-six patients with atrioventricular discordance and a variety of ventriculo-arterial connections have had appropriate intracardiac repairs. Seven of 26 patients (26.9%) died early postoperatively, but only two (11.1%) of 18 patients operated upon since May 1972 have died. Ten patients (40%) developed permanent complete heart block. Two patients died late postoperatively; 16 (84.1% of those surviving the early postoperative period) were in NYHA Class I or II at late follow-up.

Seventeen of the patients had "corrected transposition." Six developed tricuspid incompetence with regurgitation into the left atrium after repair. Two patients with atrioventricular discordance and double outlet right ventricle survived operation and are asymptomatic but have permanent pacemakers. One patient died after repair of double outlet left ventricle with atrioventricular discordance. Three of four patients with atrioventricular discordance and "pseudotruncus arteriosus" survived repair with valved external conduits and are well. One of two patients is well after repair of "isolated ventricular inversion" and ventricular septal defect.

BECAUSE OF THE RELATIVELY FEW REPORTS concerning correction of malformations with atrioventricular discordance1-4 and uncertainty as to the natural history of all the subgroups, the indications for operation in patients with these malformations have been unclear.

We have reviewed our surgical experience with patients whose malformations include atrioventricular discordance (right atrium empties through the mitral valve into left ventricle, and left atrium through the tricuspid valve into right ventricle). Various ventriculo-arterial connections occur, including that found in "corrected transposition of the great arteries."** We have grouped these various malformations with atrioventricular discordance together because of important anatomic and surgical considerations common to all. A tendency toward heart block and tricuspid valve incompetence occurs as part of their natural history, and these develop as a complication of operation with greater frequency than after repair of similar defects in hearts with atrioventricular concordance. Abnormal morphology and location of specialized conduction tissue seem responsible for the increased incidence of heart block, and the co-existence of varying degrees of Ebstein's malformation for tricuspid valve incompetence.

Material and Methods

From January 1, 1967 to October 1, 1975, 26 patients whose cardiac malformations included atrioventricular discordance underwent intracardiac repair. Patients whose atrioventricular valves open into one ventricle (commonly called single or common ventricle) are not included in this category.

Seventeen patients had "corrected transposition of the great arteries" (discordant atrioventricular connection and discordant ventriculo-arterial connection with left ventricle emptying into pulmonary artery and right ventricle into aorta10). Their ages at operation ranged from 15 months to 53 years. All of them had large ventricular septal defects, and eight also had pulmonary stenosis, either infundibular, valvular or supravalvular, or a combination of these. One of these had previously undergone pulmonary valvotomy. Three patients had tricuspid valve incompetence, and in all three a portion of the valve was displaced into the right, systemic, ventricle (Ebstein's malformation). The abdominal viscera and atria were in situs inversus in one patient, but the heart was to the left (levoposition11). Five patients had situs solitus of the viscera and atria and the heart to the right (dextroversion). Two patients had two superior vena cavae, and one a patent ductus arteriosus. Two patients had atrioventricular discordance and double outlet right (systemic) ventricle.6 Neither had dextroversion. Both had ventricular septal defect. One of these had pulmonary stenosis, and had a functioning superior vena cava-right pulmonary artery (Glenn) anastomosis. He also had stenosis at the origin of the left pulmonary artery. Double outlet left ventricle was present in one patient with atrioventricular discordance and atrial septal defect, multiple ventricular septal defects, valvular pulmonic stenosis and a previous Glenn anastomosis. "Pseudotruncus arteriosus" (common, or single, arterial trunk with ventricular septal defect, pulmonary atresia and confluent right and left pulmonary arteries) was present in four of the patients with atrioventricular discordance. Two had previously undergone Blalock-Taussig operations, and one a Pott's anastomosis. Two of these patients had situs inversus of abdominal viscera and atria and dextrocardia, and one patient had situs solitus of viscera and atria and dextroversion. Two patients had "isolated ventricular inversion" (atrioventricular discordance and ventriculo-arterial concordance). Both had ventricular septal defect. One had situs inversus of abdominal viscera and atria with dextrocardia. The other had situs solitus of the viscera and atria and the heart to the right (dextroversion). This patient also had anomalous pulmonary venous connection of the right pulmonary veins to the right atrium, corrected at operation.

Twenty-five of 26 patients were in sinus rhythm preoperatively. Six of these had first degree heart block. One patient had atrioventricular dissociation, with a ventricular rate of 46 beats/min.

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All operations were done with cardiopulmonary bypass, using standard techniques as previously described by us.\textsuperscript{12} The average time of cardiopulmonary bypass was 101 min (ranging from 55 to 193 min). Fourteen of the 27 operations (one of the 26 patients had two operations five years apart) were done at 28° to 32°C and repeated short (5 to 15 min) periods of aortic cross-clamping, and 13 were done with profound cardiac cooling and one longer period of aortic cross-clamping (averaging 37.6 min and ranging from 19 to 55 min). In this latter group, four patients had also several short (less than 15 min) periods of aortic cross-clamping after the initial longer one.

Intracardiac electrophysiologic mapping techniques were utilized in four patients,\textsuperscript{15, 16} all with “corrected transposition.” We intended to use this technique in another patient with “corrected transposition,” but she developed atrioventricular dissociation before open cardiomyotomy making electrophysiologic identification of the A-V conduction tissue impossible.

Postoperative care was standard. When atrioventricular dissociation was present, ventricular pacing was initiated before discontinuing cardiopulmonary bypass, and continued postoperatively. Two patients required re-entry early postoperatively for bleeding.

The history, operative, angiographic, and electrocardiographic findings in all patients, and autopsy material when available, were reviewed. For the purposes of this study, atrioventricular dissociation was considered present when the atria and ventricles were activated by separate spontaneous pacemakers and neither antegrade nor retrograde atrioventricular conduction was possible. The accuracy of diagnostic classification was verified for each patient. Follow-up information has been obtained on all surviving patients, in 11 through correspondence and in eight through re-evaluation at this center.

Results

Early Results

Seven of 26 patients (26.9%) who underwent operations for malformations with atrioventricular discordance died within 30 days after operation. The mortality of the subgroups of patients is shown in table 1. One 31-year-old patient, with a valved external conduit used as part of the repair in “corrected transposition,” ventricular septal defect, and pulmonic stenosis, died 17 days postoperatively as a result of sepsis. A 20-year-old man with “corrected transposition,” ventricular septal defect, and valvular and supravalvular pulmonary stenosis died 30 hours postoperatively; a few hours after operation he began to have seizures, and then became comatose. At the autopsy the repair was found to be intact. The operation was done in 1967 when a disc oxygenator was being used, which we now believe has a great tendency to produce gaseous and particulate emboli. The other five deaths occurred 1 to 96 hours after operation from low cardiac output. Four of the five patients had autopsies, and the repair was found to be complete and intact in all. No deaths seemed to be the result of the sudden development of heart block or arrhythmia.

Three deaths occurred among the 14 instances in which repeated short periods of aortic cross-clamping were used; four occurred among the 13 with profound cardiac cooling, two being in the group of four patients that also had several short periods of cross-clamping after the initial longer ischemic period.

Thirteen of the 25 patients with sinus rhythm preoperatively left the operating room with atrioventricular dissociation, the arrhythmia having developed before cardiopulmonary bypass in two, and during repair in 11 patients. The rhythm of three reverted to sinus rhythm early postoperatively. The other ten patients (40% of the initial 25 patients) either died in atrioventricular dissociation (five patients) or were discharged with it (five patients) (table 2).

The latter five had received permanent implanted pacemakers. Atrial fibrillation developed during operation in one patient (4%) and was sustained throughout the postoperative period. In addition to the three patients reverting to sinus rhythm from atrioventricular dissociation early postoperatively, two patients reverted to sinus rhythm from an A-V junctional rhythm which developed during repair; one of these still had first degree heart block and bradycardia, and the other had a bundle branch block pattern in the electrocardiogram. Nine patients remained in sinus rhythm during and after operation. Thus, 14 (56%) of the patients with sinus rhythm preoperatively manifested that rhythm at the time of hospital dismissal or death. Two of the six patients with first degree heart block preoperatively (included in the above statistics) continued in this rhythm postoperatively, one developed atrioventricular dissociation during thoracotomy, and three developed atrioventricular dissociation during repair. One of the four developing atrioventricular dissociation reverted to sinus rhythm early postoperatively.

Two of 14 patients in sinus rhythm postoperatively died, while five of ten who developed permanent atrioventricular dissociation died ($P < 0.06$).

The largest subgroup was composed of 17 patients with “corrected transposition of the great arteries” (table 3). In all, a large ventricular septal defect was closed. Three

### Table 1. Intracardiac Repair for Malformations with Atrioventricular Discordance (1967–Oct. 1, 1975)*

<table>
<thead>
<tr>
<th>Subgroups</th>
<th>Number</th>
<th>Hospital deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>“Corrected transposition”</td>
<td>17</td>
<td>4          23.5%</td>
</tr>
<tr>
<td>Double outlet LV</td>
<td>2</td>
<td>0          0.0%</td>
</tr>
<tr>
<td>Double outlet LV</td>
<td>1</td>
<td>1         100.0%</td>
</tr>
<tr>
<td>“Pseudotruncus arteriosus”</td>
<td>4</td>
<td>1          25.0%</td>
</tr>
<tr>
<td>“Isolated ventricular inversion”</td>
<td>2</td>
<td>1          50.0%</td>
</tr>
<tr>
<td>Total</td>
<td>26</td>
<td>7          26.9%</td>
</tr>
</tbody>
</table>

*2(11.1%) of 18 patients operated upon since May 1972 died. Abbreviations: RV = right ventricle; LV = left ventricle.

### Table 2. Cardiac Rhythm after Repair of Malformations with Atrioventricular Discordance (1967 to Oct. 1, 1975)

<table>
<thead>
<tr>
<th>Final rhythm*</th>
<th>Total group (26)</th>
<th>“Corr transp” (17)</th>
</tr>
</thead>
<tbody>
<tr>
<td>No.</td>
<td>%</td>
<td>No.</td>
</tr>
<tr>
<td>Sinus</td>
<td>14</td>
<td>56.0</td>
</tr>
<tr>
<td>A-V Dissoc</td>
<td>10</td>
<td>40.0</td>
</tr>
<tr>
<td>AF</td>
<td>1</td>
<td>4.0</td>
</tr>
</tbody>
</table>

*At death or hospital dismissal.

†Percent of those with sinus rhythm preoperatively.

Abbreviations: A-V = atrioventricular; Dissoc = dissociation; AF = atrial fibrillation; “Corr transp” = corrected transposition.
patients underwent replacement of an incompetent tricuspid valve at the time of repair of the ventricular septal defect, in two instances with a Starr-Edwards ball valve prosthesis, and in one a stent-mounted porcine heterograft. The tricuspid valve was found to be displaced into the right (systemic) ventricle as in Ebstein's malformation in all three patients. One of these three patients died early postoperatively. Two patients had repair of ventricular septal defect and pulmonary valvotomy, one of whom died; another patient had, in addition, infundibular resection. Repair of ventricular septal defect and insertion of a valved external conduit between left (pulmonary) ventricle and pulmonary artery for pulmonary stenosis were performed in four patients, one of whom died.

The incidence of permanent atrioventricular dissociation postoperatively in patients with "corrected transposition" is the same as in the entire group of patients with atrioventricular discordance (table 3). Six (42.9%) of 14 patients without tricuspid incompetence preoperatively developed it immediately after repair. This was identified by palpating the regurgitant jet into the left atrium, and subsequently by the usual clinical and in two instances, angiographic criteria. We believe that in no patient were chordae or leaflet tissue of the tricuspid valve damaged by the repair. This was verified at the time of valve replacement five years later in one patient. A second patient must soon undergo valve replacement.

The ventricular septal defect was repaired through the right atrium without detachment or incision of the mitral valve in 11 patients and through the left (pulmonary) ventricle in six. Among the former group, two patients (18%) died and four (36%) developed permanent atrioventricular dissociation. Among the latter two (33%) died and two (33%) developed permanent atrioventricular dissociation.

Two patients with atrioventricular discordance and double outlet right ventricle underwent operation. In one, the ventricular septal defect was repaired so that venous blood passed from left ventricle across the defect to the pulmonary artery. The anatomy did not permit this in the other patient so after closing the defect and closing the origin of the pulmonary artery from the right ventricle, a valved external conduit was placed between left ventricle and pulmonary arteries. This patient also had take-down of a previously made Glenn anastomosis, with one of the limbs of the Y-shaped conduit going then to the distal right pulmonary artery. Both of these patients developed permanent atrioventricular dissociation. Both survived and continue to be well without evident tricuspid valve incompetence.

One patient was operated upon for double outlet left ventricle and atrioventricular dissociation, with valvular pulmonic stenosis, atrial septal defect, multiple ventricular septal defects, and a Glenn anastomosis. Heart block developed during operation. The patient died of low cardiac output 24 hours postoperatively.

Four patients with "pseudotruncus arteriosus" and atrioventricular discordance underwent repair of the ventricular septal defect and insertion of a valved external conduit between left ventricle and pulmonary arteries. Two also had closure of a previously made Blalock-Taussig anastomosis, one of whom died four days postoperatively. One patient had repair of a Potts' anastomosis.

Two patients with "islated ventricular inversion" underwent repair of ventricular septal defect and intra-atrial transposition of venous return. One patient developed atrioventricular dissociation during operation and was maintained on ventricular pacing. He died 48 hours postoperatively from low cardiac output.

Late Results

Two patients, both with "corrected transposition," died after the early postoperative period. One, who also had Klippel-Feil syndrome, underwent only repair of ventricular septal defect at age 10 years. Preoperatively she had severe pulmonary hypertension which persisted after repair because of severe pulmonary vascular disease. She died five months postoperatively. The other patient was 16 years old, and underwent closure of an unusually large ventricular septal defect and insertion of a valved external conduit for pulmonary stenosis. The decision for operation was made with knowledge that the right (systemic) ventricle was abnormally small. Heart block developed at operation, and a permanent pacemaker was inserted 13 days postoperatively which functioned well during the remainder of his life. He was in congestive heart failure during most of the time after hospital dismissal, which worsened and caused death five months postoperatively. Tricuspid incompetence was present after repair, although not before, and may have contributed to the heart failure.

No patient has developed atrioventricular dissociation late postoperatively. No patient other than noted above has required reoperation.

The functional class of the patients at last follow-up is shown in table 4.

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TABLE 3. Intracardiac Repair in "Corrected Transposition of the Great Arteries" (1967-October 1, 1975)*

<table>
<thead>
<tr>
<th>Operation</th>
<th>Total</th>
<th>Hospital deaths</th>
<th>Late deaths</th>
<th>A-V dissoc.</th>
<th>Postop TVI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Repair VSD</td>
<td>7</td>
<td>1</td>
<td>1</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>Repair VSD and replace tricuspid valve</td>
<td>3</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Repair VSD and PS</td>
<td>3</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1†</td>
</tr>
<tr>
<td>Repair VSD and insertion of valved external conduit for PS</td>
<td>4</td>
<td>1</td>
<td>1</td>
<td>3</td>
<td>2</td>
</tr>
</tbody>
</table>

*1 death (8.3%) and 4 instances (33.0%) of permanent atrioventricular dissociation have occurred among 12 patients operated upon since May, 1972.
†Had valve replacement with Bjork-Shiley prosthesis 5 years after the initial operation.
§Percent of the 16 patients with sinus rhythm preoperatively.
¶Percent of the 14 patients without tricuspid valve incompetence preoperatively.
Abbreviations: VSD = ventricular septal defect; PS = pulmonary stenosis; A-V Dissoc. = atrioventricular dissociation; TVI = tricuspid valve incompetence.
Table 4. Late Results after Intracardiac Repair for Malformations with Atrioventricular Discordance (1967–Oct. 1, 1975)

<table>
<thead>
<tr>
<th>Subgroups</th>
<th>NYHA functional class</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>I</td>
</tr>
<tr>
<td>&quot;Corrected transposition&quot;</td>
<td>7</td>
</tr>
<tr>
<td>Double outlet RV</td>
<td>—</td>
</tr>
<tr>
<td>&quot;Pseudotruncus arteriosus&quot;</td>
<td>3</td>
</tr>
<tr>
<td>&quot;Isolated ventricular inversion&quot;</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>11(57.9%)</td>
</tr>
</tbody>
</table>

*Follow-up 3-56 months, average 24 months. Of 5 patients in class II are asymptomatic and taking no medications but have pacemakers.
Abbreviations: RV = right ventricle.

Discussion

Clear and well-defined terminology is essential to discussions of complex congenital heart disease. The actual names applied to malformations are not important, only their clear definition. Since certain phrases that have been understood for a long time to refer to a particular kind of malformation contain words which are not morphologically or functionally accurate or precise, we have retained them, after clearly defining them and distinguishing them by using quotation marks. The rest of the terminology which we employ is basically an outgrowth and modification of Van Praagh’s segmental approach.

Patients whose cardiac malformations include atrioventricular discordance should be treated with an awareness of the special surgical implications of atrioventricular discordance; namely, what are the ventriculo-arterial connections or the associated defects. The right ventricle relative to the left is usually on the side opposite to that occupied by the liver, but bizarre spatial relations of the ventricles may be present. Anomalies of positioning of the heart are common and complicate the surgical procedure. Differentiation of patients with atrioventricular discordance from those with single or primitive ventricle may be difficult in some instances. The incidence of atrioventricular dissociation or heart block, postoperatively, has been considerably higher than that following repair of malformations with atrioventricular concordance. We believe this high incidence is due to the tendency of the atrioventricular node and specialized conducting tissue to be in unusual locations when the atrioventricular relations are discordant. This has been described morphologically by Lev and more recently by Anderson for "corrected transposition," and electrophysiologically by Waldo and his colleagues. Abnormal morphology of conduction tissue has also been demonstrated by Anderson in some types of single, or primitive ventricle.

The risk to life of intracardiac operations in patients with "corrected transposition" is now about 10%. This is less than pertained earlier, but it suggests that the risk of repair of isolated ventricular septal defect or ventricular septal defect with mild or moderate pulmonary stenosis is greater than when there is atrioventricular discordance than might be expected. Repair of the more complex malformations with atrioventricular discordance also appears to be accompanied by somewhat higher hospital mortality rates than those pertaining to analogous patients with atrioventricular concordance.

Permanent atrioventricular dissociation has developed at operation in 40% of our patients. Knowing that atrioventricular dissociation develops as a part of the natural history of some patients with atrioventricular discordance, and that in nearly 10% of our patients it developed during surgery but before starting the repair, we believe that the incidence of its development as a permanent complication of repair will always be higher than after repair of similar lesions in hearts with atrioventricular concordance. Because the specialized conducting tissue is likely to be in an unusual position, with the atrioventricular node being in an anterior position, we believe that each stitch for the repair of a ventricular septal defect should be placed with the heart beating so as to permit electrocardiographic and visual monitoring of its possible effect on conduction. Short periods of aortic cross-clamping, with the heart and perfusate at 28°–32° C, allows this under reasonable operating conditions. If atrioventricular dissociation occurs, the stitch should be removed and replaced. Our experience with electrophysiologic mapping techniques for identification of the specialized conducting tissue before the stitches are placed is too small to allow assessment of its value in preventing atrioventricular dissociation. When atrioventricular dissociation does occur, sequential atrioventricular pacing should be used early postoperatively to provide an atrial contribution to ventricular filling.

Significant tricuspid valve incompetence has been estimated to develop as part of the natural history of the lesion in about 25% of patients with "corrected transposition," and was present in three of our 17 patients preoperatively. The techniques and risks of replacing the tricuspid valve in these patients seem to be similar to that for replacing the mitral valve in patients with concordant atrioventricular relationship.

We do not understand the mechanism of the development of tricuspid incompetence immediately after ventricular septal defect repair in six of 14 patients with "corrected transposition" who were not identified as having incompetence preoperatively. In the one patient in whom the tricuspid valve has been replaced late postoperatively, Ebstein’s malformation of the valve was present. Perhaps many of these patients have Ebstein’s malformation without incompetence which somehow develops into tricuspid incompetence after repair of the ventricular septal defect.

The pulmonic stenosis in "corrected transposition" is often subvalvar and fibrous, and specialized conduction tissue may be in close proximity to it. Usually no conus is present beneath the pulmonary valve, and this valve is in close relation to the corresponding atrioventricular (mitral)
valve. When the stenosis is severe, a bypassing valved external conduit between left ventricle and pulmonary artery must usually be added to the valvotomy. Valvotomy alone may suffice when the stenosis is mild or moderate, in which case the shunt is bidirectional, pulmonary blood flow the same or greater than systemic flow, and the patient only mildly cyanotic or acyanotic.

Patients who survive the postoperative period generally have a good result for as long as we have followed them, except for those who preoperatively had severe pulmonary vascular disease or severely underdeveloped right (systemic) ventricle or developed significant tricuspid incompetence after repair. Otherwise, even those with permanently implanted pacemakers have done well.

Acknowledgment

We express our appreciation of the patience and editorial skills of Ms. Sandy O'Brien.

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