Surgical repair of lesions associated with corrected transposition

Late results

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SUMMARY Between 1970 and 1980 19 patients aged 13 months to 47 years (mean 16 years) had surgical repair of lesions associated with corrected transposition. Four had had previous palliative surgery. Operations were performed for ventricular septal defects in 17 (multiple in two), and in addition 10 had relief of pulmonary stenosis, three had atrial septal defects closed, and three had valve replacement for left atrioventricular valve regurgitation. Two patients had annuloplasty for isolated left atrioventricular valve regurgitation.

There was a high operative mortality (37%). Twelve survivors left hospital and were followed up for three to eight years. There is concern about the high incidence of left atrioventricular valve regurgitation and progressive postoperative left sided ventricular dysfunction. Heart block after surgical intervention contributes to this, and careful pacemaker policies are necessary as two late deaths were related to this. Only one patient is asymptomatic and without complications 40 months after operation.

These disappointing late results are partly related to the onset of heart block, but it seems that independent systemic ventricular function may deteriorate in some patients. Thus surgical treatment of lesions associated with corrected transposition should be recommended only in those with important symptoms or signs of changing systemic ventricular dysfunction.

The pathological anatomy of corrected transposition was clearly described by Rokitansky in 1875,1 and the clinical features of the condition were recognised later. Corrected transposition refers to a cardiac anomaly where there are discordant atrioventricular and ventriculoarterial connections.2,3 This results in the systemic venous blood entering a right atrium which drains through a bicuspid mitral valve into a left ventricle which ejects blood into a posterior pulmonary artery. Pulmonary venous blood returns to a left atrium which empties through a tricuspid valve into a morphologically right ventricle that ejects its contents into an anterior aorta. Thus the systemic venous blood is delivered to the lungs and the pulmonary venous return to the systemic circulation so that the double anatomical discordance is corrected physiologically (or haemodynamically). The ventricle perfusing the systemic circulation is designed to support a low pressure system and its inlet valve, even when competent, has the morphology of a relatively flimsy tricuspid valve. Corrected transposition is often associated with certain intracardiac defects—namely interventricular septal defect, pulmonary valve or subpulmonary stenosis, atrial septal defect, and left atrioventricular valve regurgitation. The surgery of this condition is related to the haemodynamic disturbances which result from the presence of one or more of these conditions.

We report the surgical experience and late results in a consecutive series of patients with corrected transposition operated on in the National Heart Hospital from 1970 to 1980.

Patients and methods

Between 1970 and 1980 19 patients with corrected transposition (15 males and four females aged 13 months to 47 years (mean 16 years)) had surgical
repair of associated lesions at the National Heart Hospital (Table 1). Four had had previous palliative surgery, and two patients (cases 7 and 16), aged 41 and 20 years, had been treated before for bacterial endocarditis, which had attacked the pulmonary valve in one (case 16) and an unknown site in the other (case 7). Seventeen patients had ventricular septal defects, which were multiple muscular defects in two. In the other 15 the ventricular septal defect was muscular in one, subaortic in two, subpulmonary in one, submitral in one, inlet in one, and difficult to determine from angiographic or surgical description in nine, though it was probably subvalvular. Ten patients with ventricular septal defect had right ventricular outflow tract obstruction (gradient 53–100 mm Hg), which was at pulmonary valve level in six, subvalve in two, at valvar, subvalvar, and supravalvar levels in one, and unspecified in one.

Six patients had left atrioventricular valve regurgitation, which in five required surgical intervention at the first operation. In two patients Ebstein type malformation was the only haemodynamically important lesion, and in the other three with serious left atrioventricular valve regurgitation there was an associated ventricular septal defect. One of these three had an Ebstein malformation and one, age 47 years, had a heavily calcified valve. Before operation 11 patients were in sinus rhythm, three had intermittent or established supraventricular arrhythmias, and five had complete heart block.

Patients with the following indications were selected for surgery: heart failure with cardiomegaly resulting from a large ventricular septal defect in three patients; hypoxia (or cyanosis) due to shunt reversal as a result of severe pulmonary or subpulmonary stenosis in 12; and left heart failure resulting in pulmonary venous congestion secondary to severe left atrioventricular valve regurgitation in four cases.

All patients had disabling symptoms (grade II–IV New York Heart Association classification).6

**SURGICAL TECHNIQUE**

The surgical approach was made through a midsternal incision. Cardiopulmonary bypass was established for 50–200 minutes, mean 108 minutes, with cooling in nine patients to 26–20°C. In 11 patients transatrial repair of the ventricular septal defect was performed, through a right ventriculotomy in five patients and through bilateral ventriculotomies in one patient. Pulmonary outflow tract obstruction was relieved by means of valve bearing Dacron conduits in two patients, incorporating a Carpentier-Edwards xenograft in one patient and a fresh aortic homograft in the other. Six patients had open pulmonary valvotomy, one with patch relief of supravalvar stenosis and also relief of subpulmonary stenosis, and in a further two patients the valve obstruction was split with a bougie from below. In two patients (cases 11 and 12) isolated left atrioventricular valve regurgitation was repaired with a De Vega type annuloplasty, and in the other three patients with severe left atrioventricular valve regurgitation, which was not an isolated lesion, the valve was replaced by a dura mater valve in one (case 7), Carpentier-Edwards 31 mm xenograft in one (case...
SURVIVORS

Twelve survivors aged 22 months to 47 years left the hospital after operation, and these were followed for three to eight years.

LATE DEATHS

Four late deaths (cases 2, 7, 8, and 11) occurred 15 months to four years after the first operation. One patient (case 2) died four years after operation following bronchoscopy for haemoptysis. At necropsy the trachea and bronchial tree were normal and the cardiac anomalies were those found at operation with cardiomegaly and a patent atrial septal defect. Cardiac catheterisation 33 months previously had shown a patent foramen ovale, intact ventricular septum, severe left atrioventricular valve regurgitation, and right ventricular outflow tract obstruction of 55 mm Hg (preoperative value was 100 mm Hg). He had remained in sinus rhythm after operation.

Another patient (case 7) who presented at 41 years of age with left heart failure after the onset of atrial fibrillation had left atrioventricular valve regurgitation, pulmonary hypertension, and a small ventricular septal defect. After closure of the ventricular septal defect and replacement of the left atrioventricular valve by a dura mater valve he had recurrent atrial fibrillation.

Results

PERIOPERATIVE DEATHS

There were seven perioperative deaths. Six resulted from low cardiac output due to various and combined factors (Table 3) and the seventh was due to status epilepticus caused by septic meningitis resulting from staphylococcal septicaemia. This developed after temporary transvenous endocardial pacing for congenital heart block and heart failure. At necropsy the surgical repair of the ventricular septal defect and relief of pulmonary stenosis was perfect.

14), and by 3M Starr-Edwards prosthesis in one (case 18).

Survivors who left hospital were followed up for three to eight years (Table 2). Regular outpatient assessment was made with full history, detailed examination, and routine resting 12 lead electrocardiogram, chest x-ray examination, M mode echocardiogram, and, later, cross-sectional echocardiography. Postoperative cardiac catheterisation was performed in 10 survivors at one to 36 months; one adult survivor has refused postoperative investigation and one is alive and working in Poland three years later. Postmortem data were obtained in five cases.

Table 2 Operative technique and postoperative course in 12 survivors

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age at operation (years)</th>
<th>Operation</th>
<th>Rhythm</th>
<th>Catheter time from operation (months)</th>
<th>Length of follow up</th>
<th>LAV regurgitation</th>
<th>Reoperation</th>
<th>Id</th>
</tr>
</thead>
<tbody>
<tr>
<td>2</td>
<td>17</td>
<td>Closure VSD, pulmonary valvotomy</td>
<td>SR</td>
<td>SR</td>
<td>14</td>
<td>47</td>
<td>Nil</td>
<td>Gross</td>
</tr>
<tr>
<td>3</td>
<td>14</td>
<td>Closure VSD, pulmonary valvotomy</td>
<td>SR</td>
<td>CHB</td>
<td>6</td>
<td>86</td>
<td>Nil</td>
<td>Severe</td>
</tr>
<tr>
<td>7</td>
<td>41</td>
<td>Closure VSD, LAVVR (dura mater)</td>
<td>SR/AF</td>
<td>AF/SVT</td>
<td>1</td>
<td>15</td>
<td>Moderate</td>
<td>Mild</td>
</tr>
<tr>
<td>8</td>
<td>8</td>
<td>Closure VSD, pulmonary valvotomy</td>
<td>SR</td>
<td>CHB</td>
<td>36</td>
<td>97</td>
<td>Mild</td>
<td>Moderate</td>
</tr>
<tr>
<td>11</td>
<td>37</td>
<td>LAVV annuloplasty</td>
<td>CHB</td>
<td>CHB</td>
<td>10</td>
<td>24</td>
<td>Severe</td>
<td>Gross</td>
</tr>
<tr>
<td>12</td>
<td>32</td>
<td>LAVV annuloplasty</td>
<td>SR/AF</td>
<td>AF</td>
<td>6</td>
<td>53</td>
<td>Severe</td>
<td>Mild</td>
</tr>
<tr>
<td>14</td>
<td>47</td>
<td>Closure VSD, pulmonary valvotomy</td>
<td>AF</td>
<td>CHB</td>
<td>—</td>
<td>61</td>
<td>Severe</td>
<td>Mild</td>
</tr>
<tr>
<td>15</td>
<td>20</td>
<td>Closure VSD, LAVVR xenograft</td>
<td>SR</td>
<td>CHB</td>
<td>3</td>
<td>42</td>
<td>Nil</td>
<td>Severe</td>
</tr>
<tr>
<td>16</td>
<td>20</td>
<td>Closure VSD, closure ASD, RVOT reconstruction</td>
<td>SR</td>
<td>SR</td>
<td>12</td>
<td>40</td>
<td>Nil</td>
<td>Nil</td>
</tr>
<tr>
<td>17</td>
<td>17</td>
<td>Closure VSD, closure ASD, patch relief of supravalvar PS</td>
<td>SR</td>
<td>CHB/SR</td>
<td>—</td>
<td>36</td>
<td>Nil</td>
<td>Nil</td>
</tr>
<tr>
<td>19</td>
<td>3</td>
<td>Closure VSD, pulmonary valvotomy</td>
<td>SR</td>
<td>CHB</td>
<td>14</td>
<td>84</td>
<td>Nil</td>
<td>Moderate</td>
</tr>
</tbody>
</table>

AF, atrial fibrillation; ASD, atrial septal defect; CHB, complete heart block; LAVV, left atrioventricular valve; LAVVR, left atrioventricular replacement; PS, pulmonary stenosis; RVOT, right ventricular outflow tract; SR, sinus rhythm; SVT, supraventricular tachycardia; VSD, ventricular defect.
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A further patient (case 11) died three days after generator replacement of his permanent pacemaker, 24 months after operation. He had been paced for congenital complete heart block after the operation. The left atrioventricular valve had been repaired but moderate left atrioventricular valve regurgitation persisted together with deteriorating systemic ventricular function. At necropsy there was gross cardiomegaly, and examination of the left atrioventricular valve showed a suture straddling the valve orifice and the valve was obviously regurgitant. Ventricular tachycardia induced by the pacemaker could not be excluded as a cause of death.

The fourth late death (case 8) occurred seven years after operation during generator change of a permanent epicardial pacing system under general anaesthesia. The generator was positioned in the abdominal wall, and after wound closure, including diathermy, there was sudden progressive deterioration in systemic ventricular function which did not respond to full resuscitative procedures despite emergency thoracotomy and internal cardiac massage. Before this tragedy the child was symptomatically well with medical treatment, although postoperative catheterisation had shown severely impaired systemic ventricular function and left atrioventricular valve regurgitation.

**COMPLICATIONS IN SURVIVORS**

New and troublesome rhythm disturbances occurred in nine of the 12 survivors who left hospital. Heart block developed at operation in six patients (cases 3, 8, 14, 15, 17, and 19) and required repeated generator changes, ending fatally in one (case 8). One 37 year old patient (case 11) had congenital heart block before operation and was permanently paced. He developed complex ventricular arrhythmias and severe congestive cardiac failure due to systemic ventricular dysfunction and left atrioventricular valve regurgitation.

One patient (case 14), aged 47 years, who had established atrial fibrillation before operation, developed complete heart block at operation and required pacing. Two patients (cases 7 and 12) aged 41 and 32 at operation had preoperative intermittent atrial fibrillation and one (case 7) developed complex supraventricular arrhythmias. Sudden death occurred 15 months after operation. Three survivors aged eight to 20 years at operation maintained sinus rhythm for 40 to 57 months. Appreciable left atrioventricular valve regurgitation was present in 10 survivors and was progressive in seven (Fig). Complete heart block requiring permanent pacing was present in six of these patients, being of recent origin in five and congenital in the sixth. Two patients with atrial fibrillation had some residual left atrioventricular valve regurgitation after valve replacement. Two of three patients in sinus rhythm developed gross left atrioventricular valve regurgitation, confirmed by cardiac catheterisation six and 12 months after operation; the third patient in sinus rhythm maintained a competent left atrioventricular valve.

Seven patients had serious left atrioventricular valve regurgitation, which was not present before operation in four. One patient (case 15) underwent valve replacement four months after closure of his ventricular septal defect, and he is alive three and a half years later. One patient (case 2) died four years after operation following bronchoscopy for haemoptysis, and two patients remain symptomatically stable (cases 3 and 8).

Of the three patients in whom left atrioventricular valve regurgitation progressed seriously after operation, one awaits valve replacement (case 10) and two have died (cases 8 and 11) from pacemaker or rhythm problems. Both had poor systemic ventricular function. Left atrioventricular valve regurgitation was diminished after valve replacement in two patients (cases 7 and 14) and after repair in one other (case 12). Two survivors (cases 16 and 17) without left atrioventricular valve regurgitation do not have any residual haemodynamic lesion. One (case 17) has a demand permanent pacemaker for early postoperative complete heart block but has subsequently been in intermittent sinus rhythm.

**Fig.** Preoperative and postoperative left atrioventricular valve regurgitation in 12 survivors.
The condition of the 12 survivors three to eight years after operation is as follows: five have permanent pacemakers implanted for complete heart block, of whom three have recent or progressive left atrioventricular valve regurgitation. One of these patients required valve replacement. Two of three patients who were in atrial fibrillation before operation have mild left atrioventricular valve regurgitation after annuloplasty (case 12) or valve replacement (case 14). Two other survivors have remained in sinus rhythm but one (case 10) has developed progressive left atrioventricular valve regurgitation and is awaiting valve replacement nearly five years after the first operation. The other survivor (aged 20 years at operation) is in sinus rhythm and remains symptomatically well and has no left atrioventricular valve regurgitation. Previously he had hypoxia associated with ventricular septal defect and pulmonary valve stenosis.

Discussion

Operative mortality in this series partly reflects the learning period in dealing with complex anomalies and is similar to the experience of others. Improvement has followed a clear understanding of the position of the conducting tissue in this cardiac abnormality and with techniques of myocardial preservation. There has, however, been little or no reduction in cardiopulmonary bypass time (50–200 minutes, average 108 minutes), nor in this relatively small experience has the approach to the ventricular septal defect influenced the outcome as has been suggested. The factors predisposing to operative death, which were sometimes interrelated, were production of complete heart block, multiple ventricular septal defects, and failure to relieve adequately right sided ventricular obstruction. The site of the conduct-
Surgical repair of lesions associated with corrected transposition

Our current management of patients with corrected transposition includes surgical repair. Our approach is based on the recognition that surgical repair is indicated in patients with symptoms or signs of right atrioventricular valve regurgitation. The criteria for surgical repair are outlined in our previous publication. In the absence of symptoms or signs of right atrioventricular valve regurgitation, we recommend regular follow-up and periodic assessment of cardiac function.

We recently reported the results of surgical repair in two patients with corrected transposition and concomitant valve disease. The patients were followed for 12 and 18 months, respectively. Both patients showed improvement in their clinical status and cardiac function after surgery. We believe that surgical repair is a viable option in patients with corrected transposition who have right atrioventricular valve regurgitation.

References

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