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 Rokitsky 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 morphological 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 interatrial 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

Table 1 Clinical details of 19 patients with corrected transposition having surgical repair of associated lesions

Case No.	Sex	Previous operation	Age (years)	Age at operation (years)	Grade of symptoms*	Rhythm	Associated lesions
1	M			11	II	SR	VSD
2	M			17	III	SR	VSD PS
3	F	Left Bl	2	14	II	SR	VSD PS
4	M			3	II	SR	VSD
5	M			13 months	IV	CHB	VSD PS
6	M	Right Glenn operation	8	17	III	SR	VSD PS LAVV reg
		Left Bl	14				
7	M			41	III	SR/AF	VSD LAVV reg
8	M			22 months	IV	SR	VSD
9	M			2	II	CHB	VSD
10	F			8	II	SR	VSD PS
11	M			37	II	CHB	Ebstein LAVV reg
12	M			32	II	SR/AF	Ebstein LAVV reg
13	F			15	II	CHB	VSD
14	M			47	III	AF	VSD PS LAVV reg
15	M	Banding of PA	13	20	II	SR	VSD PS ASD
16	M			20	II	SR	VSD PS ASD
17	F	Right Bl	10	17	II	SR	VSD PS ASD
18	M			17	II	CHB	VSD
							Ebstein LAVV reg
19	M			3	II	SR	VSD PS

AF, atrial fibrillation; ASD, atrial septal defect; Bl, Blalock-Taussig shunt; CHB, complete heart block; LAVV reg, left atrioventricular valve regurgitation; PA, pulmonary artery; PS, pulmonary stenosis; SR, sinus rhythm; VSD, ventricular septal defect.

*New York Heart Association classification.

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,^{4,5} though it was probably submitral. 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 supravalar 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 supravalar 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,⁷ 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

Table 2 Operative technique and postoperative course in 12 survivors

Case No.	Age at operation (years)	Operation	Rhythm		Catheter time from operation (months)	Length of follow up	LAV regurgitation		Reoperation
			Before operation	After operation			Before operation	After operation	
2	17	Closure VSD, pulmonary valvotomy	SR	SR	14	47	Nil	Gross	
3	14	Closure VSD, pulmonary valvotomy	SR	CHB	6	86	Nil	Severe	
7	41	Closure VSD, LAVVR (dura mater)	SR/AF	AF/SVT	1	15	Moderate	Mild	
8	22/12	Closure VSD, pulmonary valvotomy	SR	CHB	36	97	Mild	Moderate	
10	8	Closure VSD, pulmonary valvotomy	SR	SR	6	57	Mild	Gross	LAVV replacement five years after operation
11	37	LAVV annuloplasty	CHB	CHB	10	24	Severe	Gross	
12	32	LAVV annuloplasty	SR/AF	AF	6	53	Severe	Mild	
14	47	Closure VSD, pulmonary valvotomy							
		LAVVR xenograft	AF	CHB	—	61	Severe	Mild	
15	20	Closure VSD	SR	CHB	3	42	Nil	Severe	LAVV replacement 4 months after operation
16	20	Closure VSD, closure ASD, RVOT reconstruction	SR	SR	12	40	Nil	Nil	
17	17	Closure VSD, closure ASD, patch relief of supra-valvar PS	SR	CHB/SR	—	36	Nil	Nil	
19	3	Closure VSD, pulmonary valvotomy	SR	CHB	14	84	Nil	Moderate	

AF, atrial fibrillation; ASD, atrial septal defect; CHB, complete heart block; LAVV, left atrioventricular valve; LAVVR, left atrioventricular valve replacement; PS, pulmonary stenosis; RVOT, right ventricular outflow tract; SR, sinus rhythm; SVT, supraventricular tachycardia; VSD, ventricular septal defect.

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. Post-mortem data were obtained in five cases.

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.

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 and complex supraventricular tachycardias, shown by electrophysiological studies. These responded to intravenous verapamil and practolol. He died suddenly 14 months after operation while taking oral verapamil, and death was assumed to be due to an arrhythmia. Postmortem examination was not performed.

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.

Table 3 Hospital deaths and late deaths in 12 survivors

Case No.	Age at operation (years)	Operation	Bypass time (minutes)	Rhythm		Residual lesion	Cause of death	Time of death from operation (months)
				Before operation	After operation			
Early deaths								
1	11	Closure of multiple VSDs	200	SR	CHB		Failure to close VSDs	
3	3	Closure of multiple VSDs	92	SR	CHB		Low output	
5	15 months	Closure of single VSD, pulmonary valvotomy	67	CHB	CHB		Septic meningitis	
6	17	Closure of single VSD, RVOT reconstruction	115	SR	CHB		Low cardiac output	
9	2	Closure of single VSD, pulmonary valvotomy		CHB	CHB		Low cardiac output, unrelieved PS	
13	15	Closure of single VSD		CHB	CHB		Low cardiac output	
18	17	Closure of single VSD LAVVR (Starr valve)	73	CHB	CHB		Low cardiac output	
Late deaths								
2	17	Closure of single VSD, pulmonary valvotomy	96	SR	SR	PS (50 mmHg), after gross LAVV regurgitation	Cardiac arrest after bronchoscopy	47
7	41	Closure of single VSD	97	SR/AF	AF/SVT	—	? Sudden arrhythmia	15
8	22 months	Closure of single VSD, pulmonary valvotomy		SR	CHB	Moderate LAVV regurgitation	Heart failure under general anaesthetic for generator change	97
11	37	LAVV annuloplasty	76	CHB	CHB	Gross LAVV	CCF + VT	24

AF, atrial fibrillation; CCF, congestive cardiac failure; CHB, complete heart block; RVOT, right ventricular outflow tract; LAVV, left atrioventricular valve; LAVVR, left atrioventricular valve replacement; PS, pulmonary stenosis; SR, sinus rhythm; SVT, supraventricular tachycardia; VSD, ventricular septal defect; VT, ventricular tachycardia.

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.^{8,9} Improvement has followed a clear understanding of the position of the conducting tissue in this cardiac abnormality and with techniques of myocardial preservation.^{2,10,11} 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.^{8,12} 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-

ing tissue in corrected transposition has been well documented.^{10 11} Our surgeons do not now consider mapping of the His bundle is necessary as it prolongs bypass and total time of operation. Electrophysiological studies in two patients before surgery and intracardiac mapping in another contributed nothing to the operation or to the outcome.^{13 14}

Our main concern in evaluating the benefits of surgery is with the clinical state of the survivors. Only one patient of the 12, who has been followed for 40 months, is without problems of rhythm or left atrioventricular valve regurgitation. This factor must be taken into consideration when radical repair is recommended for lesions associated with corrected transposition.

Progressive left atrioventricular valve regurgitation is a serious problem in the 12 survivors who left hospital because it is difficult to alleviate satisfactorily without valve replacement. Left atrioventricular valve replacement is a serious complication in children since no satisfactory biological valve is available. Imperfect valve function in the young (with mild or moderate regurgitation) is preferable to replacement with pericardial or xenograft valves.

Closing the ventricular septal defect may predispose to increasing left atrioventricular valve regurgitation by disturbing a structurally flimsy tricuspid valve, and the onset of complete heart block may cause deteriorating anatomical right systemic ventricular function contributing to left sided valve regurgitation, as with concordant right ventricular dysfunction. Deteriorating systemic ventricular function was seen in some patients after closure of the ventricular septal defect and after the onset of complete heart block, but accurate comparisons before and after operation are difficult in those patients as measurements of ejection fraction, end diastolic pressures, and angiographic appearances are all affected by changes in rhythm and the degree of left atrioventricular valve regurgitation.

Another major problem affecting the clinical course of survivors is the high incidence of rhythm disorders.^{15 16} Congenital complete heart block was present in one patient, premature complete heart block occurred in six patients, two of whom subsequently had fatal complications after revision of their pacing systems. Experienced pacing management is required in this group of patients to deal with the problems of growth, repeated generator changes, and the risk of further arrhythmias developing with age.

In the older age group malignant complex arrhythmias occurred in two patients, aged 37 and 41 years at operation. A further two patients aged 32 and 47 years at operation remain in atrial fibrillation, one with complete heart block. The learning curve of medical management has contributed in this series to dealing

with these late problems and with the early perioperative problems.

The criteria for selecting patients for radical repair of lesions associated with corrected transposition must be considered carefully. We recommend radical repair only in severely symptomatic patients with heart failure from large interventricular septal defects or appreciable left atrioventricular valve regurgitation. In hypoxic patients with severe pulmonary or subpulmonary stenosis palliation with a systemic pulmonary shunt is preferred in infancy with subsequent radical repair after adolescence. This is advisable as in most patients with severe obstruction a valved conduit is needed to prevent damage to the right coronary artery and conducting tissue and an aortic homograft may not be available. Systemic pressure in the right sided ventricle is not a direct indication for radical repair since the ventricle is designed for this. Patients not fulfilling these criteria are regularly assessed with full non-invasive investigations and subsequently referred for surgery if there is symptomatic or functional deterioration. We hope that this policy will improve the clinical state of patients with this cardiac anomaly and the prognosis of those patients who fulfil the above criteria and have surgical repair of the associated lesions.

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