LONG-TERM FOLLOW-UP (10 TO 17 YEARS) AFTER MUSTARD REPAIR FOR TRANSPOSITION OF THE GREAT ARTERIES

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Background: **The management strategies of patients who underwent Mustard repair for transposition of the great arteries were changed in the 1970s: infants became eligible for direct surgical repair, so Blalock-Hanlon atrioseptostomy could be avoided, and cold cardioplegia was introduced for myocardial preservation. Data are lacking, however, regarding whether these changes have had positive effects on the long-term outcome. We therefore conducted a follow-up study on all 91 patients who underwent a Mustard repair for transposition of the great arteries in our institution between 1973 and 1980 to assess the incidence and clinical importance of sequelae as well as health-related quality of life for these patients.** ***Methods:*** **Patients who were alive and could be traced through local registrar's offices received an invitation to participate in the follow-up study, which consisted of an interview, physical examination, echocardiography, exercise testing, and standard 12-lead and 24-hour electrocardiography.** ***Results:*** **Patients operated on in the first 4 years had a significantly higher mortality rate and higher incidence of sinus node dysfunction than did patients operated on in the subsequent 4 years (25% vs 2% and 41% vs 3%, respectively). In contrast, the incidence of baffle obstruction necessitating reoperation was significantly higher in the second group. There were no significant differences in echocardiographic findings and exercise capacity between patients operated on in the first 4 years and in the subsequent 4 years. None of the patients had right ventricular failure; a mild degree of baffle leakage or obstruction was seen in 22% of the patients, and the mean exercise capacity was decreased to 84% ± 16% of normal.** ***Conclusion:*** **The changes introduced between 1973 and 1980 have resulted in a considerable reduction of mortality and incidence of sinus node dysfunction but have also resulted in a more frequent need for reoperation. (J Thorac Cardiovasc Surg 1996;111:1158-68)**

Since the introduction of the atrial switch procedure, the outlook for patients with transposition of the great arteries has improved considerably.[1] The first follow-up results reported in 1972 by El Said and colleagues,[2] however, made clear that right ventricular failure, sinus node dysfunction, and baffle-related problems were serious drawbacks of these surgical techniques. After the introduction of the arterial switch, the atrial switch procedure was therefore gradually abandoned as the treatment of choice. Cardiologists are now seeing a large number of patients who underwent Mustard or Senning procedures with late complications, and this number will probably increase in the future. There is little information about the real incidence of complications, because long-term studies on consecutive series of patients who are actually examined at follow-up are scarce.[3, 4] Changes in surgical techniques were introduced in the 1970s to reduce the incidence of sequelae, but data are lacking regarding whether these had positive effects on the long-term outcome. We therefore conducted a follow-up study

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to evaluate the anatomic, hemodynamic, and electrophysiologic status, as well as the health-related quality of life, of all patients who underwent a Mustard repair in our institution between 1973 and 1980.

Methods

Patient selection and follow-up procedure. At our institution, in the period between 1973 and 1980, 91 patients younger than 15 years of age underwent Mustard repair for transposition of the great arteries. All operations were performed by the same surgeon. The year 1973 was chosen as starting point because the first Mustard repair was performed in our institution in that year, and 1980 was chosen as the endpoint to ensure at least 10 years of follow-up. The follow-up study started in April 1989. Patients were traced through the offices of local registrars. Eighteen patients had died and five had moved abroad. The remaining 68 patients received letters explaining the objective of the study and inviting them to participate. Fifty-eight patients (79% of the survivors, 85% of those eligible for follow-up) responded positively and took part in an extensive cardiologic examination in 1990 and 1991. Forty of these patients (69%) were male and 18 (31%) were female. The mean (± standard deviation) interval between operation and follow-up was 14.0 ± 2.1 years (range 10.1 to 17.6 years). Age at follow-up was 15.8 ± 3.9 years (range 10.4 to 27.8 years). Baseline characteristics were derived from the patient files. Examination at follow-up consisted of medical history, physical examination, standard 12-lead and 24-hour electrocardiography, exercise testing, M-mode and two-dimensional echocardiography, and pulsed, continuous-wave, and color-flow Doppler studies. The study was approved by the local Medical Ethical Review Board.

Measurements and definitions. Transposition of the great arteries was defined as "simple" if there were no concomitant abnormalities or minor abnormalities such as atrial septal defect, ventricular septal defect that did not require surgical closure, or a persisting ductus arteriosus. If the concomitant abnormality was a left ventricular outflow tract obstruction or a ventricular septal defect that required surgical closure, the transposition was defined as "complex."

Medical history. The medical history comprised a questionnaire from the Netherlands Central Bureau of Statistics that has been validated with a sample of 1510 Dutch adults younger than 35 years.[5] It uses standardized multiple-choice questions to elicit a person's assessment of his or her health status.

Echocardiographic examination. Echocardiographic examination was performed with a Toshiba SSH 160-A echocardiograph (Toshiba Medical Systems B.V., Woerden, The Netherlands). Baffle obstruction was diagnosed if there was evidence of obstruction in the two-dimensional image and the Doppler flow pattern in superior or inferior vena cava showed a pattern with a maximal antegrade flow in ventricular systole rather than diastole, which is the normal pattern after a Mustard repair.[6, 7] Obstruction to pulmonary venous drainage was diagnosed if the peak velocity of the pulmonary vein flow at the junction with the pulmonary venous atrium exceeded 1.5 m/sec. Right ventricular dimensions and contractility were judged by visual estimation of an experienced echocardiographer and a pediatric cardiologist. We did not measure these features because they are generally considered to be unquantifiable. Right ventricular dysfunction was defined to be present if there were gross dilatation and poor contractility of the right ventricle in the presence of a substantial tricuspid regurgitation and a flow velocity in the ascending aorta of less than 0.7 m/sec. The degree of tricuspid regurgitation (minimal, moderate, or severe) was estimated with color-flow Doppler scan by the width and length of the regurgitant. Pulmonary hypertension was assumed to be present if pulmonary regurgitation with a flow velocity exceeding 3.5 m/sec was found or if mitral regurgitation with a flow velocity greater than 4.0 m/sec was found in the absence of left ventricular outflow tract obstruction. Left ventricular pressure was calculated by applying the simplified Bernoulli equation to the flow velocity of the mitral regurgitation (if present) or the flow velocity in the pulmonary artery. If the velocity of this jet exceeded 3.5 m/sec and the left ventricle was not flattened but had a round shape, left ventricular pressure was considered to be elevated.

Maximal exercise capacity. Maximal exercise capacity was assessed by bicycle ergometry with stepwise increments of the workload of 20 W/min. Patients were encouraged to exercise until exhaustion. Patients were excluded from the exercise test if they could not be motivated to exercise maximally or if a concomitant factor (such as asthma, psychomotor retardation, or spastic hemiplegia) might influence the outcome of the test.

Arrhythmia. Sinus node dysfunction was defined to be present when the patient had a bradycardia-tachycardia syndrome, atrial flutter, or atrial fibrillation. The following supraventricular arrhythmias were considered to be indications of sinus node dysfunction (criteria according to Kugler[8]): sinoatrial block, a sinus arrhythmia with a beat-to-beat variation of the heart rate of more than 200%, sudden change from sinus rhythm to an escape rhythm with a frequency more than 25% lower than the sinus rhythm, nighttime bradycardia of less than 30 beats/min and daytime bradycardia of less than 40 beats/min. Ectopic ventricular activity recorded during the 24-hour electrocardiogram was considered abnormal if monoform premature ventricular contractions occurred at a rate greater than 3600 events/24 hours or if premature ventricular contractions were multiform or seen as doublets or ventricular tachycardia. Nonsustained ventricular tachycardia was defined as more than 3 but fewer than 10 consecutive beats originating from a ventricle, with a heart rate faster than 120 beats/min.

Data analysis. Because the study population was not homogeneous in terms of baseline characteristics as a result of the changes in age at operation, type of palliation before Mustard repair (Blalock-Hanlon or balloon atrioseptostomy only), and surgical techniques (complete cardiopulmonary bypass or circulatory arrest with deep hypothermia, use of cold cardioplegia) that were gradually introduced between 1973 and 1980, the population was divided into two groups. All analyses were performed for

Table I. *Baseline characteristics of 91 consecutive patients who underwent Mustard repair for transposition of the great arteries between January 1973 and December 1980*

	1973-1976 (n = 50)	*1977-1980 (n = 41)*	*1973-1980 (n = 91)*
No. of patients			
Simple TGA	31	24	55
Complex TGA	19	17	36
Age at operation (yr, ± standard deviation)			
Simple TGA	2.2 ± 2.0	0.6 ± 0.3*	1.5 ± 1.7
Complex TGA	3.6 ± 3.1	1.0 ± 1.0	2.4 ± 2.7
Preoperative palliation (n)			
No palliation	5 (10%)	3 (7%)	8 (9%)
Blalock-Hanlon atrioseptostomy	15 (30%)	0*	15 (16%)
Rashkind balloon atrioseptostomy	29 (58%)	36 (88%)	65 (71%)
Unknown	1 (2%)	2 (5%)	3 (3%)
Surgical technique (n)			
Complete CPB and hypothermia >20° C	18 (36%)	0*	18 (20%)
Circulatory arrest and hypothermia <20° C	28 (56%)	36 (88%)*	64 (70%)
Unknown	4 (8%)	5 (12%)	9 (10%)
Cold cardioplegia	4 (8%)	34 (83%)*	38 (42%)
Pacemaker implantation			
Pacemaker because of SND	7 (14%)	1 (2%)	8 (9%)
Pacemaker because complete AV-block	3 (6%)	1 (2%)	4 (4%)
Reoperation because of baffle problems (n)			
SVC-baffle obstruction	0	3 (7%)	3 (3%)
IVC-baffle obstruction	0	1 (2%)	1 (1%)
Baffle leakage	0	3 (7%)	3 (3%)
Obstruction pulmonary venous return	0	2 (5%)	2 (2%)
Reoperation for other indications (n)			
LVOT obstruction resection	0	1 (2%)	1 (1%)
LV-AP homograft	0	1 (2%)	1 (1%)
Arterial switch	1 (2%)	1 (2%)	2 (2%)
Resection aortic coarctation	0	1 (2%)	1 (1%)
Residual VSD closure	0	2 (5%)	2 (2%)
PDA closure	1 (2%)	0	1 (1%)
Interventional catheterization (n)			
VCS-baffle obstruction	1 (2%)	0	1 (1%)
Mortality (n)	15 (30%)	3 (7%)*	18 (20%)
Early (<30 days after operation)	3 (6%)	2 (5%)	5 (5%)
Late (>30 days after operation)	12 (24%)	1 (2%)*	13 (14%)

TGA, Transposition of the great arteries; *CPB,* cardiopulmonary bypass; *SND,* sinus node dysfunction; *AV,* atrioventricular; *SVC,* superior vena cava; *IVC,* inferior vena cava; *LVOT,* left ventricular outflow tract; *LV-AP,* left ventricle to pulmonary artery; *VSD,* ventricular septal defect; *PDA,* persisting ductus arteriosus.

*Significant difference between 1973 to 1976 and 1977 to 1980.

the total group of patients and separately for patients operated on in the first 4 years (1973 to 1976) and patients operated on in the subsequent 4 years (1977 to 1980). Survival was calculated according to Kaplan-Meier analysis. The χ^2 test and Fisher's Exact Test were used for the comparison of discrete variables and Student's t-test to compare normally distributed continuous variables in two groups. In the presence of a non-Gaussian distribution, the Mann-Whitney rank-sum test was used. Comparison of continuous variables in more than two groups was performed with analysis of variance. All values are expressed with their mean value ± standard deviation unless otherwise indicated. In all analyses, the level of significance was chosen at $p < 0.05$.

Results

Baseline characteristics. The baseline characteristics of the total group of 91 patients are shown in Table I. Significant differences between the first and 4-year second periods are indicated in this table. Between the 58 patients who participated in the follow-up study and the 15 patients who were known to be alive but did not participate, there were no significant differences with respect to morphologic diagnosis (simple vs complex transposition), type of palliation before Mustard repair, year of operation, and age at the time of surgical repair.

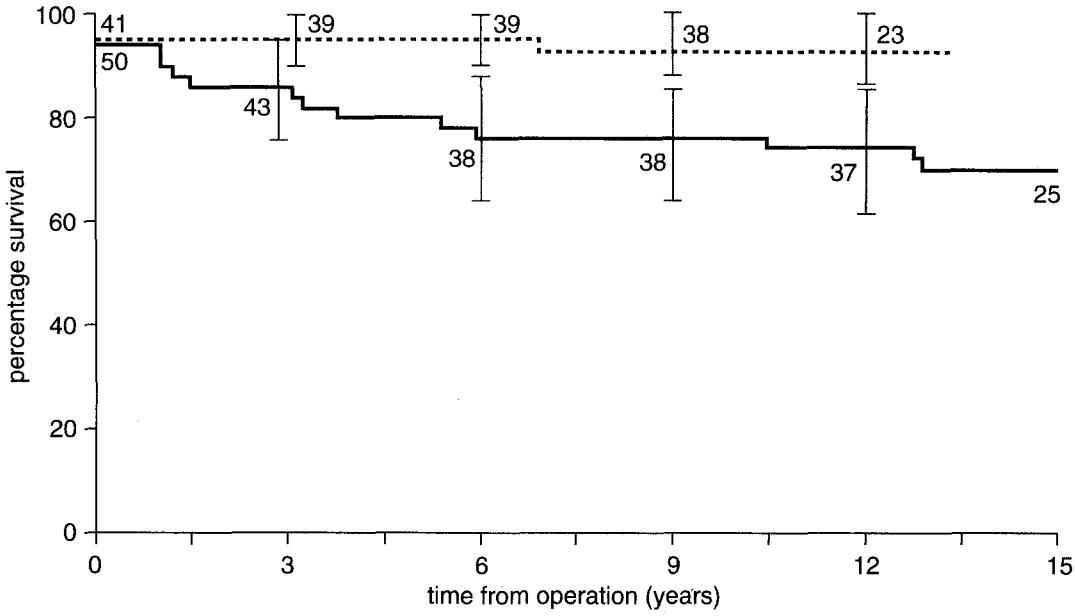

Fig. 1. Fifteen-year survival curves after Mustard operation. *Dashed line* represents patients operated on between 1977 and 1980; *solid line* represents patients operated on between 1973 and 1976.

Clinical course until follow-up

Survival. The survival to 15 years after the Mustard repair is shown in Fig. 1. Median interval between operation and follow-up was 13.9 years. The survival of patients operated on between 1977 and 1980 was significantly better than that of patients operated on between 1973 and 1976. The survival of patients with a simple transposition was not significantly better than that of patients with a complex transposition ($p = 0.8$). The mean interval between Mustard repair and late death was 5.4 ± 4.4 years (range 1.1 to 12.9 years).

The causes of death of patients who died late after operation were known for six patients: three had right ventricular failure and pulmonary edema diagnosed directly after operation, and three others had atrial flutter or fibrillation. One of these patients had, at least at last check, a properly functioning pacemaker implanted for sinus node dysfunction. Seven other patients, all operated on between 1973 and 1976, died suddenly late after operation. The cause of death could not be retrieved; none of these patients had previously documented arrhythmia or major anatomic sequelae.

Reoperation. Fig. 2 shows the 15 year event-free survival; death and reoperation were defined as events. Types of surgical reintervention are listed in Table I. The interval between Mustard operation and pacemaker implantation was 7.5 ± 3.6 years (range 2.7 to 13.8 years). The interval between Mustard repair and reoperation was 6.7 ± 4.0 years (range 1.9 to 11.3 years). Indication for pacemaker implantation was either treatment of a very low escape rhythm (as a result of sinus node dysfunction or a complete atrioventricular block) or provision of a backup in patients who needed antiarrhythmic medication for symptomatic tachycardia. Indication for an arterial switch after Mustard repair was pulmonary venous obstruction that had led to elevated left ventricular systolic pressure in one patient and a large residual nonrestrictive ventricular septal defect, which also resulted in an elevated left ventricular systolic pressure, in another patient. Reoperation was not associated with mortality.

Current health status

History. All 58 patients who participated in the follow-up study were asked to describe their health status (Table II). Forty-four patients (77%) reported that they were fatigued sooner at exercise than their peers. This proportion is significantly higher than in the general population (13%; $p < 0.05$). Six patients (11%) were taking antiarrhythmic medication (digoxin in 2 patients, propranolol, verapamil, quinidine, amiodarone). There were no differences among patients with an "excellent," "good," and "fair" personal health assessment with

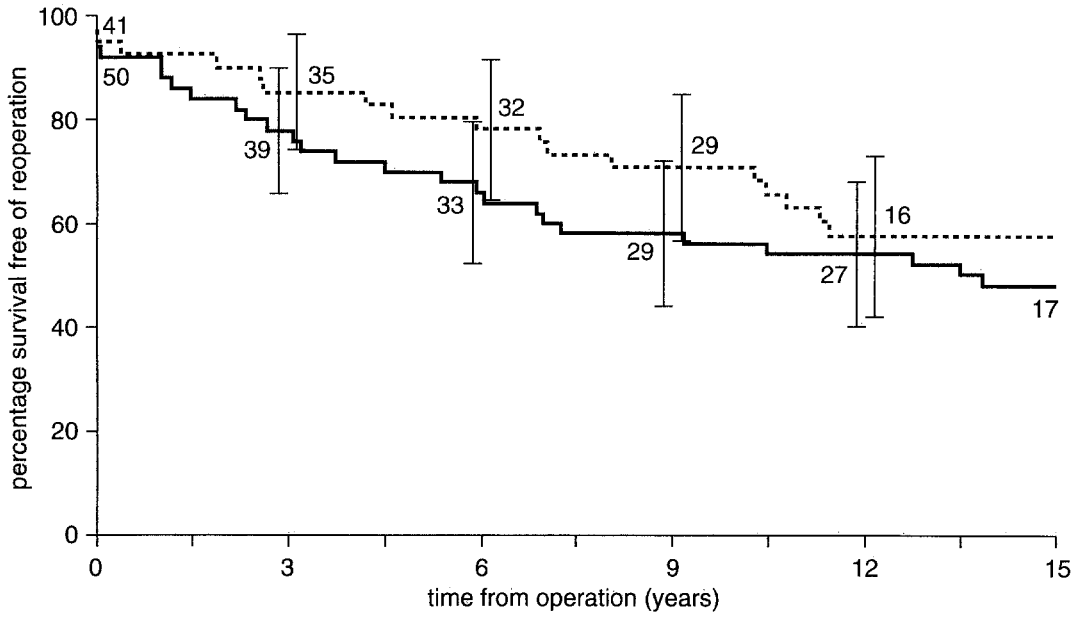

Fig. 2. Fifteen-year event-free survival curves after Mustard operation. Death and reoperation were defined as events. *Dashed line* represents patients operated on between 1977 and 1980; *solid line* represents patients operated on between 1973 and 1976.

Table II. *Personal health assessment of 58 patients long-term after Mustard repair for transposition of the great arteries, compared with a sample of 1510 persons of the normal Dutch population younger than 35 years*

	Mustard patients		*Normal population*	
	No.	*%*	*No.*	*%*
Excellent	12	21	611	40*
Good	39	67	755	50*
Fair	7	12	127	9
Not good	0	0	17	1
Bad	0	0	0	0

*Significant difference between Mustard patients and normal population.

respect to year of operation (1973 to 1976 vs 1977 to 1980), age at initial surgical repair, and presence of a simple or complex transposition.

Physical examination. The mean values of length and body weight were within the normal range for the normal Dutch population (Figs. 3 and 4). Six patients (10%) had moderate to severe chest deformity and scoliosis. Two patients had a spastic hemiplegia, and four patients had psychomotor retardation.

Echocardiography. The findings are listed in Table III. None of the patients had signs of right ventricular dysfunction. There were no differences between simple and complex transposition with respect to type and incidence of sequelae, except for elevated left ventricular pressure. Two patients with pulmonary hypertension originally had a complex transposition with large ventricular septal defect. Nine patients had a fixed subpulmonary stenosis consisting of a long fibromuscular tunnel. Of these, seven originally had a complex transposition with subpulmonary stenosis and two originally had a simple transposition with a documented normal left ventricular outflow tract before operation and without a ventricular septal defect. Three of 13 patients who had signs of baffle obstruction or leakage at follow-up had been reoperated on in the past because of baffle-related problems.

Bicycle ergometry. Six patients were excluded from the test because of spastic hemiplegia ($n = 2$) or psychomotor retardation ($n = 4$). One patient refused the test. Fifty-one patients exercised to maximum effort. The maximal exercise capacity in these patients was 84% ± 16% (range 50% to 125%) of predicted value.

There were no significant differences in exercise capacity between several subgroups that were analyzed separately: patients with "normal Mustard

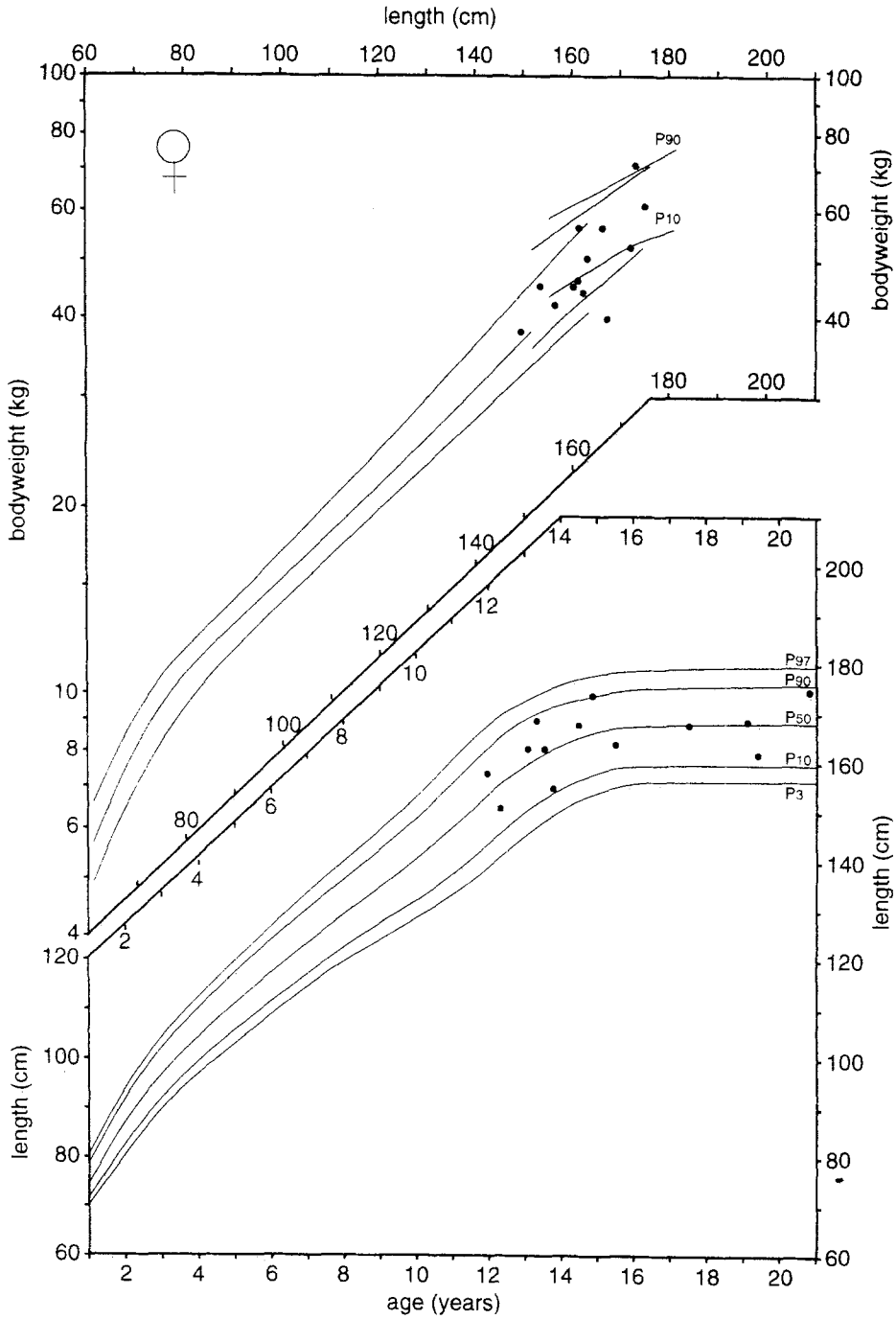

Fig. 3. Values for height and body weight for female patients at follow-up after Mustard operation. *P* represents percentile, with P_{10} being the 10th percentile.

anatomy" versus patients with echocardiographic evidence of anatomic or hemodynamic sequelae, patients with signs of sinus node dysfunction versus patients without, patients with antiarrhythmic medication versus patients without, patients with simple transposition versus patients with complex transposition, patients operated on before 1977 versus patients operated on afterwards, and patients younger than 1 year at the time of the operation versus those older than 1 year.

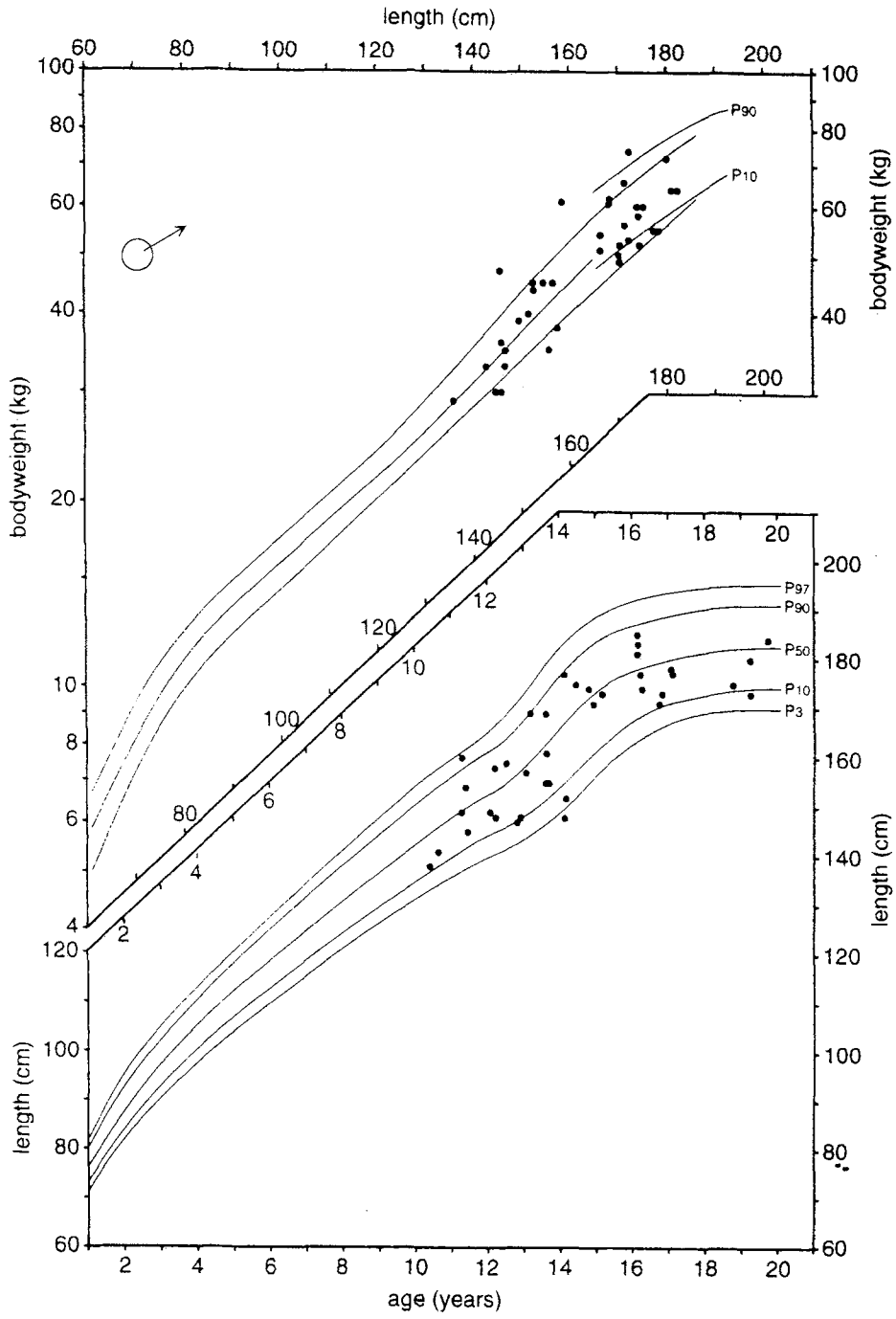

Fig. 4. Values for height and body weight for male patients at follow-up after Mustard operation. *P* represents percentile, with P_{10} being 10th percentile.

The patients who judged their health to be "excellent" had a maximal exercise capacity of 94% ± 15% of predicted, the patients who judged it to be "good" had a maximal exercise capacity of 81% ± 19%, and those who judged their health to be "fair" had a maximal exercise capacity of 71% ± 16%. These differences are significant ($p = 0.02$). Patients who reported early fatigue at exercise had a significantly lower exercise capacity than did patients who judged their exercise capacity to be normal. None of

Table III. *Echocardiography results for 58 patients at follow-up*

	1973-1976 (n = 27)		*1977-1980 (n = 31)*		*1973-1980 (n = 58)*	
	No.	*%*	*No.*	*%*	*No.*	*%*
"Normal Mustard anatomy"	12	44	17	55	29	50
Right ventricular failure	0	0	0	0	0	0
Baffle-related problems	6	22	7	23	13	22
SVC-baffle obstruction	3	11	3	10	6	10
CIV-baffle leakage	1	4	2	6	3	5
CIV-baffle obstruction	0	0	1	3	1	2
Pulmonary venous obstruction	2	7	1	3	3	5
Elevated LV pressure	7	26	4	13	11	19
Pulmonary hypertension	2	7	0	0	2	3
Fixed subpulmonary stenosis	5	19	4	13	9	16
Tricuspid regurgitation	15	56	21	68	36	62
Moderate	14	52	21	68	35	60
Severe	1	4	0	0	1	2
Aortic regurgitation	4	15	4	13	8	14
Moderate	4	15	4	13	8	14
Severe	0	0	0	0	0	0
Miscellaneous	3	11	3	10	6	10
VSD	2	7	0	0	2	3
PDA	0	0	2	6	2	3
Normal heart after arterial switch	1	4	1	3	2	3

SVC, Superior vena cava; *IVC,* inferior vena cava; *LV,* left ventricular; *VSD,* ventricular septal defect; *PDA,* patent ductus arteriosus.

the patients had increased ventricular ectopic activity during or directly after the exercise test.

Standard 12-lead electrocardiography. A narrow QRS pattern was seen in 49 patients (86%), 47 of whom had signs of right ventricular dominance. The two patients who underwent arterial switch operation after Mustard repair had completely normal electrocardiograms. Eight patients had a wide QRS-pattern: five had a right bundle branch block and three had a left bundle branch block. Thirty-five patients (60%) had a regular sinus rhythm, eight patients (14%) had a stable atrial rhythm, four patients (7%) had a stable junctional rhythm, and three patients (5%) had a supraventricular rhythm with a wandering pacemaker. Of the remaining eight patients, seven (12%) had a pacemaker rhythm and one (2%) had an atrial flutter with 3:1 conduction. The mean PR interval was 0.16 ± 0.03 seconds (range 0.08 to 0.26 seconds). None of the 10 patients with echocardiographic evidence of elevated left ventricular pressure had electrocardiographic signs of increased left ventricular activity. There were no electrocardiographic differences between the group of patients who originally had simple transposition of the great arteries and the group with complex transposition.

Twenty-four hour electrocardiography. The results of the 24-hour electrocardiogram, which could be obtained for all 58 patients, are shown in Table IV. Eight of these 58 patients had a pacemaker implanted in the past. Seven had predominantly pacemaker rhythm. The remaining patient, who had a pacemaker (and was taking digoxin and propranolol) because of sinus node dysfunction, had no pacemaker activity during the 24-hour electrocardiogram but had continuous atrial flutter. The 24-hour electrocardiograms of the six patients who used antiarrhythmic medication (two of whom had pacemakers to support the antiarrhythmic treatment) showed atrial flutter in two patients, bradycardia-tachycardia in one patient, a continuous pacemaker rhythm in one patient, and no arrhythmia in two patients.

The total number of surviving patients with sinus node dysfunction (either asymptomatic or treated with a pacemaker) was 12 (21%), 11 of whom were operated on between 1973 and 1976. Four of 12 patients who had a pacemaker implanted in the past were not evaluated at follow-up: one patient had died suddenly (possibly of arrhythmia) despite a well-functioning pacemaker, one patient had gone abroad, and two patients refused to participate in the follow-up study.

There were no differences in the incidence of ventricular or supraventricular arrhythmia between patients with simple and complex transpositions or

Table IV. *Results of 24-hour electrocardiographic registration of 58 patients at follow-up*

	1973-1976 (n = 27)		*1977-1980 (n = 31)*		*1973-1980 (n = 58)*	
	No.	*%*	*No.*	*%*	*No.*	*%*
No arrhythmia	1	4	14	45*	15	26
Supraventricular arrhythmia	17	65	6	19*	23	40
Atrial flutter	2	7	0	0	2	4
Bradycardia-tachycardia syndrome	5	19	0	0	5	9
Other signs of SND	10	37	6	19	16	28
Ventricular arrhythmia	15	58	10	32	25	43
VT >10 beats	0	0	0	0	0	0
VT 3–10 beats	3	11	1	3	4	7
PVC doublets	2	7	1	3	3	5
Multiform PVCs	10	37	8	26	18	31
AV block	1	4	1	3	2	2
Complete	1	4	0	0	1	2
Second-degree Mobitz type	0	0	1	4	1	2
Pacemaker rhythm	5	19	2	6	7	12
Originally SND	4	15	1	3	5	9
AV block	1	4	1	3	2	4

SND, Sinus node dysfunction; *VT,* ventricular tachycardia; *PVC,* premature ventricular complex; *AV,* atrioventricular.
*Significant difference between 1973–1976 and 1977–1980.

between patients with "normal Mustard anatomy" and patients with anatomic or hemodynamic sequelae. Atrial flutter or bradycardia-tachycardia syndrome was seen in five of the nine patients who had a Blalock-Hanlon atrioseptostomy before Mustard repair and in two of the 17 patients who had a Rashkind procedure in the same period ($p = 0.02$). At multivariate analysis, none of the possible risk factors for sinus node dysfunction (older age at the time of the surgical correction, no cold cardioplegia, Blalock-Hanlon atrioseptostomy) reached statistical significance.

Discussion

There is a striking difference in outcome between patients operated on before 1977 and those operated on afterward. In the early group, both late mortality (25%) and incidence of sinus node dysfunction (41%) were significantly higher than in the late group (2% and 3%, respectively). It is unlikely that these differences are the result of the differences in interval after operation because the mean interval between Mustard repair and event (death or pacemaker implantation for sinus node dysfunction) was significantly shorter than even the minimum duration of follow-up for the late group. In contrast to others,[9, 10] we therefore conclude that the better outcome in the late group is the positive net effect of management changes designed to prevent sinus node dysfunction and sudden death. It is likely that older age at the time of the operation (longer duration of severe hypoxemia, which could affect the myocardium), scarring of the atrium (Blalock-Hanlon atrioseptostomy), limited myocardial preservation during operation (no cold cardioplegia), and lesser experience of the surgeon all contributed to the greater rate of events in the early group. Multivariate analysis, however, failed to single out a single statistically significant independent determinant. We agree with others[3, 11] that the prognosis for patients with a Mustard-type correction is not necessarily as dim as expected on the basis of earlier data.[12]

However, despite the apparent good survival and the low incidence of sinus node dysfunction in the group of patients operated on since 1977, it is evident that there remains ample reason for concern for most patients that cardiologists will see in daily practice because many patients were operated on in the earlier years of cardiac surgery.

Arrhythmia. Although not entirely absent in the late group, the problem of sinus node dysfunction is almost completely confined to the early group of patients. This is probably a reflection of the state of the art of cardiac surgery in the early years, before improvements had been implemented.

It is noteworthy that one patient died suddenly, probably of arrhythmia, despite the presence of a pacemaker. This finding is in agreement with reports of others that implantation of a pacemaker does not totally prevent sudden death,[10] which is probably

caused by ventricular fibrillation, whether induced by supraventricular arrhythmia or not. Because of the association between arrhythmia and late death, it seems justified to approach these arrhythmias more aggressively; implantable antitachyarrhythmic devices should be considered in selected patients.

Indications of sinus node dysfunction according to Kugler[8] were often present in the early group (37%) but were also found in the late group (19%). The interpretation of these arrhythmias detected on a surface electrocardiogram is hazardous, and one can only speculate on the causative mechanism and clinical relevance. On the one hand, they can be considered normal because they have been reported to occur also in the normal population.[13-15] On the other hand, it is known from electrophysiologic studies that sinus node recovery time is abnormal in as many as 84% of patients after Mustard repair.[16, 17] It is therefore possible that these subtle signs of sinus node dysfunction represent real damage to the sinus node. An already damaged sinus node might be more vulnerable to the changes as a result of aging, such as loss of number of sinus node muscle cells[18, 19] and deterioration of sinus node function,[20, 21] which explains an elevated incidence of arrhythmia in "normal" older people. Clearly, longer follow-up is needed.

Anatomic and hemodynamic sequelae. The clinical importance of most of the anatomic and hemodynamic sequelae appears to be limited. These sequelae were associated neither with decreased exercise capacity (compared with patients without sequelae), nor with complaints, nor with increased incidence of arrhythmia. None of the sequelae that were detected at follow-up resulted in reoperation. Three patients (5%) had serious sequelae that were not amenable to repair: two patients had pulmonary hypertension and one patient had severe tricuspid regurgitation.

Reoperation. Reoperations for baffle leakage or obstruction were confined to the late group. The technical difficulty of creating an adequate sized baffle in the smaller children operated on during this period is probably the cause of these problems.[22] There was no mortality associated with reoperation, but three of these patients had echocardiographic evidence of residual baffle problems at follow-up. We therefore conclude that reoperation for baffle obstruction or leakage can be performed safely but is not always successful from a functional point of view.

Exercise capacity. Exercise capacity was equally diminished in both early and late group. Apparently, the changes that led to improved survival and a lower incidence of sinus node dysfunction have not improved exercise capacity. The cause of decreased exercise capacity after Mustard repair remains a subject of controversy. It is even questioned whether decreased exercise capacity is inherent in the Mustard type repair.[23-26] We believe that decreased exercise capacity is the result of the Mustard repair and is not the result of a flaw in the exercise study protocol because we found completely normal exercise capacity (with the same test protocol and normal values) in patients of the same age group after surgical closure of atrial septal defect or ventricular septal defect.[27, 28] This corresponds with the perception of the patients themselves that they fatigue earlier than others. The fact that exercise capacity corresponds well with patients' health self-assessment underlines the fact that exercise capacity is an important determinant of health-related quality of life.

Right ventricular capability. The ability of the right ventricle to function as the systemic ventricle for a lifetime is the last concern. In contrast to others, we did not find right ventricular failure in any of the survivors.[29] Others have demonstrated with serial measurements that right ventricular performance did not deteriorate through the years.[30] Questions about long-term right ventricular function nevertheless remain, and only longer duration of follow-up of this specific patient group can answer them.

We conclude that the incidences of sinus node dysfunction and late sudden death are high in the total group of patients who underwent Mustard-type repair in our institution between 1973 and 1980. The changes in management strategies that have been introduced during this period, however, have resulted in considerable reduction in incidence of these late complications. This change makes the long-term outlook for patients better probably than previously believed on the basis of earlier data, but the frequent need for reoperation and the reduced exercise capacity show that the Mustard repair continues to have restrictions, even for these patients.

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