Palliative Mustard operation for transposition of the great arteries: late results after 15-20 years

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Abstract

Objective—To assess the clinical, functional state, and complications late (15–20 years) after palliative Mustard operation.

Design—Examination and evaluation of all patients presenting in adolescence and adult life after palliative Mustard operation for transposition of the great arteries and pulmonary vascular disease.

Setting—Grown-up Congenital Heart Unit specialised in the care of adolescents and adults with congenital heart disease, designated as having “quaternary” status within a tertiary referral centre for cardiac diseases.

Patients and methods—Database searched for patients referred after palliative Mustard for classic transposition of the great arteries. Ten patients aged 18–31 (mean (SD) 25.9 (5.2)) years with a palliative Mustard operation performed at age 1.7–15 (mean (SD) 9 (4.6)) years were fully evaluated by echocardiography, exercise testing, Holter monitoring, and magnetic resonance imaging or radionuclide ventriculography, or both 15–20 years later.

Results—One patient died aged 25 years with biventricular failure (ability index 3/4), haemoptysis, and atrial flutter, eight were well (ability index 2), and one disabled (ability index 3). Arterial oxygen saturation at rest was 85–98% (mean (SD) 93.8 (4.4%)) decreasing to 59–87% (mean 77 (9.5%)) after limited exercise. Symptomatic arrhythmias occurred in four patients, atrial flutter being the most common, and two had sinus node dysfunction. Significant tricuspid regurgitation occurred in four patients.

Conclusion—Patients with palliative Mustard operation have a low incidence of symptomatic ventricular dysfunction and despite exercise limitation by hypoxia, continue to live active, near normal lives until their thirties.

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Keywords: palliative Mustard operation; late results after 15–20 years; transposition of the great arteries, congenital heart disease

The concept of palliative Mustard operation was introduced by Lindesmith *et al*¹ in 1972 for the treatment of patients with transposition of the great arteries and raised pulmonary vas-

cular resistance. This is an intra-atrial baffle repair which leaves or creates² a ventricular septal defect and has been shown to give good symptomatic relief over 7–13 years.^{3–5} Knowledge about the state and fate in adults is sparse. This study reports the findings in all adults patients seen in the Grown-up Congenital Heart Unit during the past 6–23 (mean (SD) 11 (5.7)) years, 15–20 years after palliative Mustard operation.

Patients and methods

The Grown-Up Congenital Heart Unit database, established in 1976, was searched for patients with complete transposition of the great arteries and a previous palliative Mustard operation referred to a Grown-Up Congenital Heart Unit (adolescents aged 12–19 years and adults) initially at the National Heart Hospital, subsequently in the Royal Brompton Hospital. There were 10 patients (seven women and three men) aged 18–31 (mean (SD) 25.9 (5.2)) years operated on between 1973 and 1978 at the age of 1.7–15 (mean (SD) 9 (4.6)) years.

Table 1 summarises the patients' data. A large ventricular septal defect was left open in eight patients and was created at operation in two. Nine patients had increased pulmonary vascular resistance or biopsy-proven pulmonary vascular disease, or both. Three of these patients (nos 3, 8, and 10) had had previous pulmonary artery banding, but pulmonary vascular resistance was thought to be too high to safely close the ventricular septal defect at the time of the Mustard procedure. Another patient with previous pulmonary artery banding (no. 1) had normal pulmonary artery pressure and had a palliative Mustard operation because of a small right ventricle and straddling tricuspid valve.^{4,6} Although this patient did not have increased pulmonary vascular resistance like the other nine and the rationale for leaving the ventricular septal defect open in this case was different, the patient was included in this series as he underwent a palliative Mustard procedure preserving the original principles of the operation. One patient (no. 2) had had previous repair of preductal coarctation of the aorta but the patent arterial duct was left open.

The nine surviving patients were assessed in 1993, 15–20 (mean (SD) 17 (2)) years after the operation. Protocol investigation was performed in the outpatient clinic where eight patients attended regularly; one (no. 9) was investigated in another clinic and data

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Table 1 Clinical data in 10 patients with transposition of the great arteries who underwent palliative Mustard operation

Patient no	Age in 1993 (years)	Diagnosis and associated defects	Previous operations	Age (years)	Additional surgery at palliative Mustard	Age at palliative Mustard	Postoperative PAP	
							mm Hg*	% systemic
1	28	TGA, VSD, small RV, straddling TV	PAB, atrial septectomy	23/365 11/12	DePAB	11	24/12 (18)	25
2	30	TGA, IVS, preductal CoA, PDA, JAA, right aortic arch kyphoscoliosis	CoA repair	13	Create VSD	13	115/80 (90)	100
3	29	TGA, VSD	Atrial septectomy, PAB, right B-T shunt	2/12 2/12 7		10	95/50 (70)	100
4	30	TGA, VSD	Atrial septectomy	7		15	120/70 (90)	100
5	24	TGA, IVS, JAA			Create VSD	5.5	85/50 (60)	100
6	31	TGA, VSD Mild valvar PS Dextrocardia, visceral and atrial situs inversus, kyphoscoliosis	Atrial septectomy	7	Pulmonary valvotomy	13	70/30 (45)	60
7	20	TGA, VSD	Balloon atrial septostomy	4/12		2	75/40 (55)	100
8	28	TGA, VSD, Kyphoscoliosis	Atrial septectomy, PAB	3/12 1.5	DePAB	8	60/35 (48)	50
9	18	TGA, VSD, JAA				1.7	80/50 (60)	85
10 (Dead, 1990)	25	TGA, VSD	Atrial septectomy, PAB	3	DePAB	10	100/58 (70)	100

*Values in parentheses indicate mean pressures.
B-T, Blalock-Taussig; CoA, coarctation of the aorta; DePAB, debanding of the pulmonary artery; IVS, intact ventricular septum; JAA, juxtaposition of the atrial appendages; PAB, pulmonary artery banding; PAP, pulmonary artery pressure; PDA, patent ductus arteriosus; PS, pulmonary stenosis; RV, right ventricle; TGA, complete transposition of the great arteries; TV, tricuspid valve; VSD, ventricular septal defect.

obtained. The records of one patient (no. 10) who died in 1990 were reviewed. The study protocol included clinical examination, chest radiography, electrocardiography, trans-thoracic echocardiography (10 patients), exercise testing using the modified Bruce protocol (nine patients), oxygen saturation measurements at rest and on exercise (nine patients), and 24 h ambulatory electrocardiography (seven patients). Functional state was designated by the Ability Index designed by Warnes and Somerville.⁷ All patients had postoperative cardiac catheterisation between 1979 and 1993, and radionuclide ventriculography or magnetic resonance imaging, or both, for the assessment of ventricular function⁸ between 1990 and 1993. Echocardiography was used to evaluate intracardiac anatomy, intra-atrial baffle, ventricular size, and contractions; atri-ventricular valve regurgitation was graded by colour flow Doppler as mild, moderate, or severe according to the extension and width of the regurgitant jet into the atrial venous compartment. Right and left ventricular ejection fractions were measured by magnetic resonance imaging or first pass and equilibrium gated radionuclide ventriculography, or both (normal ejection fraction for right ventricle > 35%, left ventricle > 45%).

Statistical analysis was performed using the Student's *t* test for paired data and Fisher's exact test to compare differences between groups. A *p* value < 0.05 was considered significant. Results are expressed as mean (SD).

Results

FUNCTIONAL STATE

Eight patients (ability index 2) currently lead active lives. One patient (no. 2) with severe kyphoscoliosis has been classified as ability index 3 for four years since the age of 26 and another (no. 10) who died aged 25 from

chronic right heart failure was ability index 3/4 for 3 years before death.

Improvement in symptoms and signs (failure to thrive, heart failure, dyspnoea, and cyanosis) occurred initially in all patients after palliative Mustard operation and was maintained for 13.4 (4.7) years until age 15–30 (mean (SD) 22.4 (5)) years. Five patients (nos 1, 3, 4, 5, and 9) aged 18–30 years showed no deterioration in symptoms, exercise capacity, or Ability Indices during the period of observation to date. The remaining five patients (nos 2, 6–8, and 10) developed heart failure or increasing cyanosis, or both between the ages of 15 and 24 years (fig 1). Heart failure and progressive cyanosis developed in two patients (nos 2 and 10) four and twelve years after the operation with subsequent deterioration of exercise capacity and Ability Indices; both had pulmonary artery pressures at systemic level and developed recurrent haemoptyses and paroxysmal atrial flutter. Patient no. 10 died 15 years after the operation while awaiting heart lung transplantation. Patient no. 2 who is now aged 30 has the lowest exercise and functional capacity of the survivors, 17 years after the operation. Another patient (no. 8) developed paroxysmal atrial flutter nine years after palliative Mustard operation with subse-

Figure 1 Development of heart failure (HF) and cyanosis (CY) with change in Ability Index (AI) in time after palliative Mustard operation.

Table 2 Ability index, exercise performance, and systemic arterial oxygen saturations in 10 patients after palliative Mustard operation

Patient no	Age (years)	Ability index	Exercise test		Systemic arterial oxygen saturation (%)		Heart rate (beats/min)		Blood pressure (mm Hg)	
			Stage	Total duration (min and s)	At rest	At peak exercise	At rest	At peak exercise	At rest	At peak exercise
1	28	2	3	6 26	97	79	98	154	125/80	160/90
2	30	3	1	1 52	91	72	101	136	110/70	120/70
3	29	2	4	9 40	96	80	105	174	100/70	135/80
4	30	2	3	7 16	85	59	74	149	90/60	105/65
5	24	2	4	11	98	87	80	165	105/80	125/70
6	31	2	4	9 53	96	76	89	156	120/80	155/80
7	20	2	4	9 10	94	74	94	170	100/75	150/90
8	28	2	3	6 26	96	86	73	150	125/85	180/80
9	18	2			91					
10 (Dead)	24	3/4	2	4 30			99	150	120/80	130/85

Figure 2 Systemic arterial oxygen saturations before (at rest) and 15–20 years after (at rest and on exercise) palliative Mustard operation. Preop., preoperative; postop., postoperative.

quent heart failure which was controlled by digoxin and diuretics; since then, the patient has experienced no further deterioration in symptoms or functional state and her ability index remains at 2 20 years after the procedure. Cyanosis in four patients, in two (nos 4 and 9) since the operation and apparent in two (nos 6 and 7) 10·5 and 14 years later, did not progress.

Table 2 summarises the Ability Indices and exercise performance with systemic arterial oxygen saturation. Four patients (nos 3, 5, 6, and 7) (Ability Index 2) reached stage 4 of the modified Bruce protocol with systemic arterial oxygen saturation at rest of 94–98%, decreasing to 74–87% on exercise; three patients (nos 1, 4, and 8) (Ability Index 2) reached stage 3 with arterial oxygen saturation at rest of 85–97%, decreasing to 59–86%. Patient no. 2 (Ability Index 3) stopped at stage 1 with a decrease in arterial oxygen saturation from 91% to 72% and patient no. 10 with (Ability Index 3/4) stopped at stage 2. Exercise testing in these patients was carried out with caution, always supervised by a physician.

Four patients (nos 1, 3, 5, and 8) did not develop cyanosis at rest, four had mild (nos 4, 6, 7, and 9), and two had moderate cyanosis (nos 2 and 10). The mean (SD) systemic arte-

rial oxygen saturation increased significantly ($P < 0\cdot001$) from a preoperative value of 58·9 (12)% (range 45–74%) to 93·8 (4)% (range 85–98%) at rest, decreasing to 77 (9·5)% (range 59–87%) on exercise, as measured 17 (2) years after the operation (fig 2). The mean (SD) haemoglobin concentration decreased from a preoperative value of 18·6 (3) (range 15·5–23·5) g/dl to 14·9 (2·7) (range 12–18·7) g/dl 17 (2) years after the operation; this difference was not statistically significant.

One patient (no. 8) with systemic arterial oxygen saturation of 96%, pulmonary artery pressure 50% of systemic (60/35, mean 48 mm Hg), and pulmonary vascular resistance 75% of systemic (7 U.m²) had two uncomplicated pregnancies and two healthy children were delivered normally with combined management of obstetrician and cardiologist. This patient and two others (nos 5 and 6) with pulmonary artery pressures at systemic and 60% of systemic levels respectively had elective sterilization.

PULMONARY ARTERY PRESSURE

Table 1 shows the pulmonary artery pressure in patients aged 8–27·8 (mean (SD) 20·2 (6)) years measured 10 (6·3) years after palliative Mustard operation. The pulmonary artery pressure was normal (24/12, mean 18 mm Hg) in one exceptional patient (no. 1) who had had pulmonary artery banding and underwent palliative Mustard operation because of small right ventricular size and straddling tricuspid valve. In the remaining nine patients, the pulmonary artery pressure was systemic in six (nos 2, 3, 4, 5, 7, and 10), two of whom developed haemoptysis, and 50–85% of systemic in three (nos 6, 8, and 9); the mean pulmonary artery pressure being 45–90 (mean (SD) 65·3 (16)) mm Hg and the mean (SD) pulmonary vascular resistance 18·8 (12) U.m². Two of the patients with previous pulmonary artery banding had systemic pulmonary artery pressures (nos 3 and 10) and in another (no. 8) the pulmonary artery pressure was 50% of systemic. There was no change in pulmonary artery pressure or pulmonary vascular resistance after palliative Mustard operation compared with preoperative values.

VENTRICULAR FUNCTION AND ATRIOVENTRICULAR VALVE REGURGITATION The right ventricular (systemic ventricle) ejection fractions ranged from 14% to 60%

Figure 3 Development of right ventricular (RV) dysfunction, tricuspid regurgitation, and arrhythmia by age in patients after palliative Mustard operation. Regurg, regurgitation.

Figure 4 Development of right ventricular (RV) dysfunction, tricuspid regurgitation, and arrhythmia in time after palliative Mustard operation. Regurg, regurgitation.

Table 3 Resting routine and 24 h ambulatory electrocardiogram (ECG) results in 10 patients after palliative Mustard operation

Patient no	Ability index	Standard ECG	Age at onset of symptomatic arrhythmia (years)	24 h ambulatory ECG
1	2	1 degree A-V block, VEs		1 degree A-V block, intermittent 2 degree A-V block with 2:1 conduction, multifocal VEs, SVE couplets, 1 episode of VT
2	3	SR (paroxysmal atrial flutter), RBBB	17	SR, sinus bradycardia, frequent VEs (isolated, couplets, bigeminy)
3	2	SR, RBBB		
4	2	SR, RBBB		SR/occasional junctional, sinus pauses (2 s)
5	2	SR, RBBB		SR, frequent VEs (isolated, couplets, triplets, bigeminy)
6	2	SR, frequent VEs	19	SR, frequent VEs (couplets, bigeminy), few SVEs
7	2	SR, RBBB		
8	2	SR (paroxysmal atrial flutter)	17	SR, multifocal VEs (isolated, couplets, bigeminy), SVEs (isolated, couplets, bigeminy)
9	2	SR		
10 (Dead)	3/4	Atrial flutter	24	Polymorphic VT

A-V, atrioventricular; RBBB, right bundle branch block; SR, sinus rhythm; SVEs, supraventricular extrasystoles; VEs, ventricular extrasystoles; VT, ventricular tachycardia.

(mean (SD) 40.4 (14)%). Six patients (nos 1, 3, 5, 7, 8, and 10) developed impaired (< 35%) right ventricular ejection fractions by age 14.5–29 (mean (SD) 22 (5.3)) years, 11–19 (mean 14.2 (3)) years after palliative Mustard operation (figs 3 and 4) which was asymptomatic in five patients (ejection fractions 32–35%) and associated with symptoms in one (no. 10) who died 3 years later from severe right (ejection fraction 14%) and left (ejection fraction 38%) ventricular dysfunction with severe tricuspid valve regurgitation. Four patients (nos 2, 4, 6, and 9) aged 18–31 (mean 27.3 (6)) years retain normal right ventricular function (ejection fractions 51–60%).

The left ventricular (pulmonary ventricle) ejection fractions ranged from 38% to 62.5% (mean (SD) 52.2 (7)%). Among the nine survivors, none had impaired (ejection fraction < 45%) left ventricular function.

Tricuspid regurgitation had developed in seven patients by the age of 14.5–31 (mean (SD) 22.4 (5.4)), 8–18 (mean (SD) 13 (3)) years after the operation (figs 3 and 4) and was trivial in three (nos 1, 7, and 8), moderate in two (nos 4 and 5), and severe in two (nos 6 and 10), although only one was symptomatic. Tricuspid regurgitation was present in five of the six patients with impaired right ventricular ejection fraction. Two patients (nos 4 and 6) with moderate and severe tricuspid regurgitation, respectively, had good right ventricular function; tricuspid regurgitation had been present for 6 years and was not progressive in patient no. 4 (aged 30 years) while it was recently detected in patient no. 6 (aged 31). One (no. 3) of the four patients with previous pulmonary banding had mild mitral regurgitation not seen in the rest. None of the patients had obstruction in venoatrial pathways with magnetic resonance imaging or echocardiography.

ARRHYTHMIAS

Table 3 shows the current routine electrocardiogram and 24 h ambulatory electrocardiogram results. Four patients experienced

symptoms from their arrhythmia; nos 2, 8, and 10 with atrial flutter and no. 6 with frequent ventricular extrasystoles. The basic rhythm is sinus in eight patients which is maintained throughout in four and associated with recurrent atrial flutter in two (nos 2 and 8); the basic rhythm was atrial flutter in patient no. 10 who died. Two patients had sinus node dysfunction according to previously defined surface electrocardiogram/Holter criteria⁹; no. 2 with sinus bradycardia (33 beats/min during sleep) and no. 4 with sinus pauses (2 s). Although only two patients had ventricular extrasystoles on resting standard electrocardiogram, Holter monitoring revealed ventricular extrasystoles in five patients, polymorphic ventricular tachycardia in one, and supraventricular extrasystoles in three. No arrhythmias were provoked by exercise.

Discussion

This series is selected by survival and therefore may not reflect the outcome of all patients undergoing palliative Mustard procedure. The size of the original cohort of patients having palliative Mustard operations is not known as they were operated on in several hospitals, most in The Hospital for Sick Children, Great Ormond Street where 41 patients had this operation between 1973 and 1980 with 38 hospital survivors.³

The mean follow up period of 15–20, mean (SD) 17 (2) years is the longest of published reports.^{3–5} Most patients in this series (8 of 10) maintained a good Ability Index (2) and were able to work with their lives minimally disturbed by cardiac problems despite the limitation of exercise tolerance from arterial desaturation with cyanosis on exertion.

The results of this study support the findings of others reporting improved exercise tolerance, a rise in arterial oxygen saturation, a decrease in polycythaemia after the procedure up to a mean of 10.5 years^{3 4 10} and also confirm that improvement has been maintained until over 30 years in some.

The major haemodynamic result of the redirection of pulmonary and systemic venous return is the net gain in systemic arterial oxygen saturation achieved by changing streaming and reducing the amount of systemic venous blood reaching the aorta while increasing the effective pulmonary blood flow.¹¹ The situation becomes like Eisenmenger's syndrome with ventricular septal defect and it can be anticipated that the course will be similar with death from haemoptysis, right heart failure, cerebral incident, or sudden from unanticipated arrhythmia.¹² Right heart failure which occurs in Eisenmenger's syndrome usually after 40–50 years may occur earlier after palliative Mustard operation as hypoxia is established earlier in infancy and in present in childhood. As yet, there is no evidence to confirm this and the left ventricular (pulmonary ventricle) ejection fractions of the nine survivors remain within normal limits, although frank failure has occurred in one.

There has been much concern about ventricular function after the Mustard operation in view of the differences in structure and contraction pattern of the right ventricle functioning as a systemic ventricle and the deleterious effects of preoperative hypoxia on the myocardium.^{13–15} Right ventricular and less pronounced left ventricular dysfunction have been reported in asymptomatic and symptomatic patients after the Mustard operation.^{13–15} Although we found reduced right ventricular ejection fractions in six patients after palliative Mustard operation, it was mild and asymptomatic in five and severe with symptoms in only one (no. 10) who is now dead. It is interesting that progressive and symptomatic right ventricular dysfunction was not common, presumably because of the blow-off valve effect of the ventricular septal defect. The frequency of symptomatic right ventricular dysfunction (10%) is less than that reported after ordinary Mustard procedure¹⁶ (18%) in adult patients of similar age and follow up period, assessed by the same methods and criteria, but the difference is not statistically significant. Despite a high afterload due to increased pulmonary vascular resistance left ventricular function was well preserved in nine patients; and only one patient with previous pulmonary artery banding had mitral regurgitation which was mild and asymptomatic. Tricuspid regurgitation was common (seven of 10 patients; moderate in two and severe in two); this could be explained by the association of a large ventricular septal defect which distorted the anatomy. Tricuspid regurgitation was not associated with symptoms except in one patient (no. 10). Not all the patients with tricuspid regurgitation had right ventricular dysfunction (and vice versa), although they coexisted in five patients.

The seven patients who had serial Holter monitoring were found to have some form of arrhythmia, giving symptoms in four. Recurrent atrial flutter occurred in three patients and was associated with disabling symptoms. The frequency of symptomatic arrhythmia after palliative Mustard operation

is similar to that reported after the Mustard procedure.¹⁶ The mechanism of arrhythmia after palliative Mustard operation should be similar to that of Mustard operation involving a diseased sinus node and interrupted atrial pathways,¹⁷ as the intra-atrial rerouting procedure is common to both operations. Sinus node dysfunction, the most common abnormality after the Mustard operation,^{16–18–19} was less frequently detected in this series of patients with palliative Mustard (20%) compared with that reported in patients who had the Mustard procedure (59%)¹⁶ on Holter monitoring although the difference was not significant ($P = 0.057$). No patient had complete heart block, including those in whom a ventricular septal defect was created at operation. Only one patient had first and second degree atrioventricular block. Ventricular extrasystole complexes were common (five patients), giving symptoms in one. The only patient who presented with ventricular tachycardia had established ventricular failure which occurs similarly after Mustard operation and in other patients with chronic ventricular dysfunction and dilatation. In this series of patients after palliative Mustard operation, right ventricular dysfunction, tricuspid regurgitation, and arrhythmia developed by a mean (SD) age 22 (5) years, beginning around age 15 years and increasing in frequency thereafter until age 30.

The results of this study show in a small, selected group of survivors beyond childhood that palliative Mustard operation provides good palliation in the long term, although there is exercise limitation from hypoxia. Because of Eisenmenger's physiology and the added problems of a right ventricle functioning as a systemic ventricle, arrhythmia following intra-atrial baffle repair, and hypoxia established early in infancy, patients are unlikely to survive much beyond the fifth decade. Earlier and more radical treatment has mostly prevented the development of pulmonary vascular disease in transposition of the great arteries so such cases rarely appear. If such patients present, however, the palliative Mustard procedure can be considered for those in whom pulmonary vascular disease has developed,^{20–22} albeit with a limited life span but a reasonable chance of 20–30 years of improved quality of life.

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