Transposition of great arteries

Early results of Mustard's operation in paediatric patients

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Between May 1969 and June 1975, 43 patients with d-transposition of the great arteries, ranging in age from 2 months to 13 years (mean 19 months), underwent surgical correction. In the first 3 patients Mustard's original method was followed. Later a modified procedure was performed using a Teflon patch. Forty-two of the 43 patients had previously had 50 various palliative procedures, of which 30 were balloon atrial septostomy only. Five patients died in hospital (up to 5 weeks after operation). Of the 32 patients with simple repair of transposition of the great arteries (including 3 with ligation of a persistent ductus arteriosus), 3 died. One patient died as a result of complete AV block, one as a result of renal damage associated with unrelieved coarctation of the aorta, and one of cerebral infarction and peritonitis which probably were initiated pre-operatively. Six patients who had additional surgery for pulmonary stenosis survived, including 3 patients who had closure of ventricular septal defect. Of another 5 patients with closure of ventricular septal defect, including 2 who had debanding of the pulmonary artery, 2 died in a low cardiac output state. One infant operated on for subpulmonary stenosis developed permanent complete AV block and was successfully treated with a pacemaker. All the patients with combined operations had some problems postoperatively. The survivors improved greatly, but 2 patients died suddenly one, and one and a half years after the operation respectively.

Haemodynamic correction of transposition of the great arteries by redirecting the pulmonary and the systemic circulation at the atrial level is based on the pioneer work of Albert (1954), of Senning (1959), and of Johansson (1963). Mustard in 1964 described the first case successfully operated upon using a pericardial patch to redirect the caval blood to the mitral orifice and the pulmonary venous blood to the tricuspid ostium. Since then this has become a standard procedure, using either a pericardial or a Dacron patch. Accompanying significant lesions also have to be corrected during the procedure. The present report reviews our experience with this treatment in infants and children.

Procedures

Right-sided thoracotomy was used in the first patients with simple transposition of the great arteries and no previous heart operation. In

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patients with atrial septal defect after a Blalock-Hanlon procedure, with pulmonary stenosis, or with ventricular septal defect, a sternal split was used, and later this became the standard procedure in all patients. In the first 3 patients we used the original Mustard technique and sutured the patch in front of the coronary sinus. To avoid the region of the atrioventricular node and the bundle of His, in later patients the patch has been placed behind the sinus, draining its blood together with the pulmonary venous blood. In the first 3 patients pericardial patches were used. Later we have used very thin, knitted Teflon (U.S.C.I.¹ No. 3105) because it is pliable, easy to form, and does not stretch.

A standardization of the baffle size and shape according to the patient's weight was worked out, and also the suture line has been somewhat modified (Tjønneland and Hall, 1976). Ventricular septal

¹U.S. Catheter+Instrument Corp., Murray Hill, New Jersey 0794, U.S.A.

TABLE 1 *Year of correction*

	1969	-70	-71	-72	-73	-74	-June 1975	Total No.
No. of patients	1	2	3	11	16	8	2	43
No. of deaths		1		2	1	1		5

defects have been closed from the right atrium or from the left ventricle with Teflon patches. Post-operatively, the patients have been ventilated using a naso-tracheal tube for 1 to 2 days, or more if necessary. In the beginning of our series 5 patients had tracheostomies. All patients had continuous monitoring of direct arterial pressure, superior caval venous pressure, and electrocardiogram for a minimum of 1 to 2 days.

Subjects

After only 1 to 3 operations per year in the first years 1969 to 1971 the number reached a peak in 1973, because of an accumulation of patients. The lower number performed in 1974 may represent what is to be the expected annual need for this operation in Norway (Table 1). Before the procedure, 42 of the 43 patients had undergone 50 separate palliative procedures; 30 of the patients had balloon atrial septostomy only (Table 2). Blalock-Hanlon operations were done in 3 patients with an inadequate balloon septostomy, and in 6

TABLE 2 *Palliative procedures before correction*

Type of procedure	No. of patients
Balloon atrial septostomy alone	30
Balloon atrial septostomy plus	
Blalock-Hanlon	3
Pulmonary arterial banding	2
Resection of coarctation of aorta plus	1
Pulmonary arterial banding plus	
Ligation of persistent ductus arteriosus	
Blalock-Hanlon alone	4
Blalock-Hanlon plus	
Blalock-Taussig anastomosis	2
	42
Medical treatment alone	1

TABLE 3 *Haematocrit values before correction*

Haematocrit (vol. %)	40-49	50-59	60-69	70-79	80-89	Total No.
No. of patients	2	5	19	16	1	43

TABLE 4 *Age at correction*

	Months		Years			Total No.
	2-6	7-12	1-2	3-6	7-13	
No. of patients	6	6	16	11	4	43
No. of deaths	3			2		5

patients in a period before the balloon technique was introduced. Two of these had pulmonary stenosis and in them a subclavian-pulmonary anastomosis was performed as well.

All the patients were cyanotic. Most of them also had functional dyspnoea, and some had enlarged livers. The haematocrit was 60 per cent or more in 36 of the 43 patients (Table 3).

The youngest patient was 2 months old and weighed 2.9 kg at the operation. Table 4 shows that 12 of the 43 patients (28%) were operated upon in their first year of life. The mean age of all the patients was 19 months.

Twenty-four patients had transposition of the great arteries and atrial septal defect only. Five patients had additional heart lesions which were not corrected as they were considered to be of minor importance. One was the youngest patient, who had a small persistent ductus arteriosus. Four patients had a mild to moderate pulmonary subvalvar stenosis on the angiocardigram, with systolic pressure gradients ranging from 28 to 48 mmHg (3.7 to 6.4 kPa) between the left ventricle and the pulmonary artery or wedged pulmonary vein position. One of these also had a moderate coarctation of the aorta.

In 14 of the 43 patients the Mustard procedure was combined with surgery for other heart lesions (Table 5).

TABLE 5 *Associated lesions and corrective procedures*

	Total No.	Deaths
A. Mustard procedures only	29	3
B. Mustard procedures plus other correction		
Ligation of ductus arteriosus	3	
Pulmonary valvulotomy or subvalvular resection or dilatation*	3	
Closure of ventricular septal defect	3	1
Closure of ventricular septal defect + debanding	2	1
Closure of ventricular septal defect + pulmonary valvulotomy/subvalvular resection	3	
	43	5

*One patient also had a Blalock-Taussig shunt ligated.

TABLE 6 Early course after correction

	Simple	Complex
Uneventful	24	
Total atrioventricular block	1*	1
Atrial arrhythmias requiring treatment	3	4†
Ventricular fibrillation		1
transient hemiplegia		2*
Cardiogenic shock		
Bleeding + reoperation	1	
" " + renal failure	1*	
" " + atelectasis	1	
Congestive heart failure		3
Cerebral infarction + peritonitis	1*	
Total no. of patients	32 (3*)	11 (2*)

*Deaths.

†Two patients had also congestive heart failure.

Results

Five (12%) of the 43 patients died. Of the 32 patients (Table 6) who had a *simple Mustard procedure* (including the 3 with ligation of persistent ductus arteriosus) 3 died (9%). One of these developed total AV block with a ventricular rate of 90, which later fell to between 50 and 60 per minute. The use of a pacemaker might have saved him (a later patient in our series, with a combined Mustard procedure and pulmonary valvotomy plus sub-pulmonary resection also developed total AV block, but is now doing well on a radio-frequency pacemaker). A second patient died of renal failure; he had unrelieved coarctation of the aorta. A 2-month-old infant of 2.9 kg died of cerebral infarction and peritonitis, both of which were probably present before the operation; the clinical assessment was difficult as the patient was very ill, and the final diagnosis was only made at necropsy.

Of the 11 patients who had a *Mustard operation*

combined with other cardiac surgery, 2 died (18%), both of low cardiac output after closure of a ventricular septal defect. One was 8 months old and had hyperkinetic pulmonary hypertension. The other was banded at 1 month of age, and the banding was rather tight when he had the Mustard procedure 3½ years later.

Arrhythmias occurred frequently. Besides the total AV block already mentioned, the most serious were repeated episodes of ventricular fibrillation. In one case this occurred on the 4th postoperative day, followed by transient hemiparesis some days later, but with no permanent sequelae. Four patients had atrial flutter and were electrically converted. Another 2 cases had frequent ectopic atrial beats which responded to digitalis. In 3 patients with ventricular septal defect and pulmonary stenosis, severe congestive heart failure developed. They gradually improved on medical treatment.

While 24 of the 32 simple Mustard procedures made an uncomplicated recovery, all the patients with additional heart surgery had some sort of complication.

All the 38 survivors improved considerably, 30 of them being without cardiac symptoms. However, 2 of these died suddenly of no apparent cause one, and one and a half years, respectively, after the operations. Six patients have slight cyanosis clinically, possibly caused by leakage around the baffle. In 11 cases the chest x-ray film shows some increase in the heart size; this has occurred more commonly in the patients who have had complex procedures.

Discussion

In the past few years experience with the Mustard operation has been reviewed in a number of reports

TABLE 7 Results of Mustard's operation for transposition of great arteries (TGA)

References	Centre	Period in years	Total no. of patients (and mortality*)	Subgroups (and mortality)† Simple TGA
Danielsen <i>et al.</i> (1971)	Rochester, Minn., U.S.A.	1964-69	69 (25-36%)	25 (3-12%)
Waldhausen <i>et al.</i> (1971)	Philadelphia, U. S.A.	1965-70	32 (6-19%)	18 (0)
Clarkson <i>et al.</i> (1972)	Auckland, N.Z.	1965-70	56 (13-23%)	46 (7-15%)
Breckenridge <i>et al.</i> (1972)	London, U.K.	1965-71	200 (36-18%)	138 (15-11%)
Ihenacho <i>et al.</i> (1973)	Birmingham, U.K.	1968-71	37 (10-27%)	33 (7-21%)
Champsaur <i>et al.</i> (1973)	Toronto, Canada	1963-72	123 (26-21%)	87 (11-13%)
Gutgesell and McNamara (1975)	Houston, U.S.A.	1967-72	31 (8-26%)	20 (4-20%)
Stafford and McGoon (1973)	Rochester, Minn., U.S.A.	1969-72	31 (4-13%)	19 (2-10%)
Subramanian (1974)	Buffalo, N.Y., U.S.A.	1969-73	67* (14-21%)	34 (3-9%)
This series	Oslo, Norway	1969-75	43 (5-12%)	32 (3-9%)

*All were below 2 years of age and treated under deep hypothermia.

†First figure in parentheses is the number of fatalities, the second figure is the percentage. TGA, transposition of great arteries; PS, pulmonary stenosis; VSD, ventricular septal defect.

(Table 7). The results reported here compare favourably with the others. In our material the median age was lower and the proportion of complicated cases was about the same as in most of the other series.

Improved results are reported with increased experience. Early mortality in the last year of the series of Breckenridge *et al.* (1972) was only 14 per cent; it was only 15 per cent in the last 2 years of the series of Champsaur *et al.* (1973), and 10 per cent in ours.

Mustard, and most of the other authors, have used pericardium as baffle material. After having tried this first, and then Dacron, Stark *et al.* (1974) went back to pericardium, because caval obstruction occurred with Dacron patches in infants. Stafford and McGoon (1973), however, had no trouble with Dacron. We have used Teflon and we have so far had no clinical problems, but follow-up studies are needed for an adequate evaluation of the baffle.

There is no general agreement yet as to the most suitable age for the Mustard procedure. Waldhausen *et al.* (1971) and Gutgesell and McNamara (1975) prefer the second year of age for transposition of the great arteries with intact ventricular septum, with palliative procedures before that. But the former group have advocated earlier correction if the child's general condition deteriorates. A substantial mortality has been observed however after palliative treatment in the first 12 months of life (Tynan, 1971; Parsons *et al.*, 1971). More recently the trend has been to do early correction, particularly in simple transposition (Aberdeen, 1971). Neutze and Clarkson (1973) reported 2 hospital deaths (11%) after correction of 18 infants with transposition of the great arteries and intact ventricular septum. They now accept 3 months as the optimal age for correction of simple transposition

of the great arteries using deep hypothermia. Subramanian (1974) had 2 deaths in 23 infants (9%) and also preferred correction at 3 to 6 months of age. Stark *et al.* (1974), who used routine cardiopulmonary bypass, had only 2 (4%) deaths in their 52 infants with simple transposition of the great arteries.

In our series there were 12 infants, 3 of whom died; of the 9 survivors, 8 had simple transposition of the great arteries and one had resection of a subpulmonary stenosis. Of those who died only one had simple transposition of the great arteries, and his death was unrelated to cardiac surgery. Another died of renal failure, with unrelieved coarctation of the aorta, which possibly should have been repaired before the Mustard procedure. The third had a ventricular septal defect with pulmonary hypertension.

The management of complicated transposition of the great arteries is controversial, the results after the Mustard procedure often being less satisfactory. Those with large or multiple ventricular septal defects, where there may be irreversible pulmonary hypertension, have hitherto first had palliation by banding of the pulmonary artery. This treatment, at 4 to 6 months of age, was still recommended by Bonham-Carter (1973). However, it seems that the current trend is to recommend early primary correction (usually using deep hypothermia). Neutze and Clarkson (1973) found that banding did not give satisfactory palliation. They performed successful primary correction in 4 infants at 2 to 4 months of age, but in another 4 operated upon during the first month of life only 1 survived. Champsaur *et al.* (1973) found that the overall risk of banding with subsequent Mustard procedure, repair of ventricular septal defect, and debanding, was formidable, and also recommended primary correction, before 6 to 8 months of age. Subramanian (1974) had used the same approach in the presence of uncontrolled failure, but was reviewing this policy. Stark *et al.* (1974) used partly deep hypothermia, partly ordinary bypass, in correcting transposition of the great arteries associated with ventricular septal defect (TGA+VSD) with 4 out of 10 infants surviving. Of our 5 patients with TGA+VSD, 2 died, 1 at 3½ years of age (after a banding at 3 months), the other at 8 months of age, with hyperkinetic pulmonary hypertension (and no banding). Our future policy will probably be early primary correction.

Repair of ventricular septal defect and pulmonary stenosis combined with the Mustard procedure has carried a high mortality (Table 7). The procedure of Rastelli (1969) is now recommended (Danielsen *et al.*, 1971; Bonham-Carter, 1973).

GA+PS	TGA+VSD	TGA+PS+VSD	Other
(1)	29 (16-55%)	14 (5-36%)	
(0)	9 (3-33%)	3 (3)	
-	6 (4)	3 (1)	1 (1)
(1)	44 (12-27%)	14 (7-50%)	1 (1)
(2)	—	1 (0)	1 (1)
(2)	22 (10-45%)	9 (3)	
(2)	1 (1)	1 (1)	
(0)	7 (1)	3 (1)	
-	21 (4-19%)	12 (7-58%)	
(0)	5 (2)	3 (0)	

Transposition of the great arteries with ventricular septal defect and coarctation or interruption of the aorta poses even more difficult problems. Clarkson *et al.* (1972) had no survivors in this group. Stark *et al.* (1974) stressed the importance of first resecting the coarctation and banding the pulmonary artery, and had patients surviving this procedure awaiting correction. We had one such a patient who survived correction after this initial procedure.

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