

Transposition of the great arteries

A review of 37 cases after Mustard's operation

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Ihenacho, H. N. C., Patel, R. G., Singh, S. P., Astley, R., and Parsons, C. G. (1973). *Thorax*, 28, 448-452. **Transposition of the great arteries. A review of 37 cases after Mustard's operation.** Thirty-seven children with transposition of the great arteries have undergone Mustard's operation in Birmingham Children's Hospital from May 1968 to December 1971. Thirty-three of these had simple transposition of the great arteries and four were complicated. All but two had had some previous palliative procedures. Ten patients died before discharge from hospital and there were two late deaths, one after three months and the other after eight months. Bleeding, dysrhythmia, and hypotension were common immediate postoperative complications, while recurrent heart failure and persistent atrioventricular block occurred in five and four patients respectively.

Mustard's operation (Mustard, 1964; Mustard *et al.*, 1964) is now a standard procedure for the treatment of transposition of the great arteries (TGA). It is a satisfactory operation for uncomplicated cases such as transposition with atrial septal defect (Breckenridge *et al.*, 1972), offering hope for a condition which otherwise has an appalling prognosis (Leibmann, Cullum, and Bellock, 1969). Parsons, Astley, Burrows, and Singh (1971) reported the result obtained in the first 65 infants who had been treated by balloon atrial septostomy at the Children's Hospital, Birmingham, and we now report our experience with the first 37 children who have undergone Mustard's operation.

CLINICAL MATERIAL

Between May 1968 and December 1971, 37 children with TGA have undergone Mustard's operation. Twenty were males and 15 females. There were 32 cases of simple transposition and five were complicated. Two had an additional ventricular septal defect and one each, pulmonary stenosis, patent ductus arteriosus, and total anomalous pulmonary venous drainage. Teflon cloth was used for constructing the baffle in the first case, Dacron was used in the last six cases, while pericardium was used in the others. All operations were performed with cardiopulmonary bypass and moderate hypothermia.

AGE AND WEIGHT AT OPERATION The youngest patient was 6 months old and the oldest 69 months. Average

age at operation was 34.45 months. Weight at operation ranged from 5.04 kg to 18.15 kg with a mean of 11.42. The average age at operation has been reduced over the four-year period (Table I).

TABLE I
MUSTARD'S OPERATION 1968-71

Year	No. Operated	Average Age at Op. (mth)	Average Weight at Op. (kg)	Hospital Deaths	Late Deaths
1968	3	42.3	12.47	1	0
1969	6	43.67	12.37	3	0
1970	12	25.3	11.32	0	1 ¹
1971	16	26.6	9.53	6	1 ²
	37	34.46	11.42	10	2

¹ 3 months after operation.
² 8 months after operation.

PREVIOUS PALLIATIVE PROCEDURES Twenty-nine patients had balloon atrial septostomy (Rashkind and Miller, 1966) while three had operative atrial septostomy. Four had atrial septectomy (Blalock and Hanlon, 1950). Two infants did not have any palliative procedure; one had a satisfactory shunt through an atrial septal defect and the other had total anomalous venous drainage into the right atrium.

ADDITIONAL OPERATIONS Five patients had some other operation at the time of the Mustard procedure (Table II). One ligation of patent ductus arteriosus, one pulmonary valvotomy, and one correction of supracardiac total anomalous venous drainage were performed. Two patients had a repair of the ventricular septal defect through the tricuspid valve. Four

TABLE II

ADDITIONAL OPERATIONS DURING CORRECTION OF TRANSPOSITION OF THE GREAT ARTERIES

Operation	No. of Patients
Repair of ventricular septal defect	2
Pulmonary valvotomy	1
Ligation of patent ductus arteriosus	1
Correction of total anomalous pulmonary venous drainage	1
Re-exploration of wound	4

TABLE III

DURATION OF STAY IN HOSPITAL FOLLOWING MUSTARD'S OPERATION

Year	No. of Patients Surviving Mustard's Operation	Average Duration of Stay (days)
1968	2	37
1969	3	27.7
1970	12	23
1971	10	17.4
	27	26.2

had the chest re-explored for evacuation of blood clot within 24 hours of the Mustard's operation.

TOTAL DURATION OF POSTOPERATIVE STAY IN HOSPITAL One patient was fit to be discharged after six days. The majority of patients were discharged within four weeks. With increasing experience of the operation and improving postoperative care, there has been a reduction in the duration of hospital stay (Table III).

DEATHS There were 10 deaths in the immediate postoperative period. Seven of these had uncomplicated TGA, two were complicated by a ventricular septal defect, and one by total anomalous pulmonary venous drainage. Prolonged hypotension was the cause of seven deaths, four within 24 hours, two on the fourth, and one on the fifth day. One patient who died on the tenth day had evidence of severe cerebral damage demonstrated by electroencephalography. Pyopneumothorax and septicaemia caused one death on the fifteenth day while another death occurred after a stormy postoperative period of 34 days during which the patient had disturbances of rhythm—nodal rhythm and frequent ventricular ectopics, not responding to treatment.

Two late deaths occurred after satisfactory recovery from operation. One suffered transient loss of consciousness (less than a minute's duration) three months after operation, but no cause was found. Following an attack of vague abdominal pain he died suddenly. Necropsy failed to establish the cause of death. Another patient died eight months after operation, soon after com-

pletion of the cardiac catheterization which was performed as part of the investigation of recurrent cardiac failure.

POSTOPERATIVE COMPLICATIONS The following complications were recorded.

Bleeding Wound bleeding occurred commonly but in the majority of patients was slight and stopped within 24 hours. In all four cases where bleeding was excessive the chest was re-opened for evacuation of clot. One of these patients died.

Disturbance of rhythm Transient disturbances of rhythm, most commonly nodal rhythm and supraventricular tachycardia, were observed during the first few days in about one-third of our patients. Treatment was not usually required. One had alternating episodes of nodal tachycardia and atrial fibrillation soon after operation, reverting to sinus rhythm on the sixth day. In one case bradycardia was noted, especially when the patient was lying on the left side. This ceased after two days but he developed A-V nodal rhythm after the fifth day. When last seen four months later he was well and in sinus rhythm. Nodal rhythm and frequent ventricular ectopics preceded death on the thirty-fourth day in one patient while another whose A-V nodal rhythm persisted over the first week was eventually discharged well but in 2:1 block.

However, dysrhythmias in two children have been troublesome. In a boy aged 8 (case 5, Table IV) episodes of paroxysmal supraventricular tachycardia, atrial flutter, and fibrillation with a ventricular rate of 220 to 300 per minute necessitated three readmissions. Initially, these responded to intravenous administration of lignocaine and digoxin. The last episode which occurred nine months after operation was resistant to lignocaine but was terminated by intravenous propranolol. He has been maintained on oral propranolol, and the episodes of paroxysmal atrial tachycardia now occur infrequently, usually after a respiratory infection, and are of shorter duration. A 3-year-old girl (case 11, Table IV) was re-admitted with atrial tachycardia and heart failure four months after Mustard's operation. She had been having a maintenance dose of digoxin (0.05 mg twice daily orally) since operation. Sinus rhythm was restored with intravenous propranolol followed by a direct current counter-shock of 25 joules. Digoxin and diuretics were continued and when last seen she was well, in nodal rhythm with a ventricular rate of about 60 but no evidence of heart failure.

Complete atrioventricular block occurred in six patients soon after operation (Table IV). Temporary transvenous pacing was used in two of these immediately the condition was recognized, and was continued until spontaneous reversion to sinus rhythm after 48 hours. One child aged 8 years (case 3, Table IV) has required permanent pacemaking with an inductive coupled pacemaker (Abrams, Hudson, and Lightwood, 1960). Ventricular rate has remained satisfactory in three other children with atrioventricular block.

TABLE IV
DYSRHYTHMIAS AFTER MUSTARD'S OPERATION

Year	Case No.	Dysrhythmia	Onset	Rhythm in March 1972
1968	1	Paroxysmal atrial fibrillation	2 days	A-V block
	2	A-V block	3 days	A-V block
	3	A-V block	Immediate	Ventricular rate 60-70/min A-V block; permanent pacing
1969	5	Paroxysmal supra-ventricular tachycardia; atrial flutter and atrial fibrillation	4 months	Sinus rhythm maintained with pro-pranolol
	9	Atrial fibrillation; nodal rhythm	1 day	Sinus rhythm
	11	Atrial tachycardia	4 months	Nodal rhythm
	15	Bradycardia; nodal rhythm	2 days	Sinus rhythm
	22	A-V block	5 days 1 day	A-V block; ventricular rate about 90/min
1971	25	A-V block	1 day	A-V block; ventricular rate about 80/min
	28	Nodal rhythm; frequent ventricular ectopics	1 day 34 days	Died
	32	A-V block; paced for 2 days	1 day	Sinus rhythm
	37	A-V block; alternatively with nodal rhythm; 2:1 block	1 day 7 days	A-V block 2:1

Recurrent cardiac failure Five patients have had recurrent cardiac failure which was sufficiently severe to require inpatient treatment on three occasions. All five have been taking digoxin and large doses of diuretics since operation. Two have been shown to have obstruction of the systemic venous return due to contraction in the intra-atrial baffle. Details of these cases will be reported in another communication.

DISCUSSION

Mustard (1968), in a review of further experience with his operation for transposition of the great

arteries with an intact ventricular septum, recommended a minimum weight for operation of 9.0 kg. Over the years we have tended in this centre to operate on younger children and the average weight has dropped from 12.5 kg in 1968 to 9.5 kg in 1971. Children with an additional ventricular septal defect required early operation to prevent pulmonary vascular disease developing; it is often an early complication (Viles, Ongley, and Titus, 1969; Ferencz, 1966). In patients with an intact ventricular septum early operation is designed to avert the risk of intravascular thrombosis to which these children are exposed between the time of septostomy and corrective surgery (Parsons *et al.*, 1971). Nine of our children weighed less than 9 kg and five died; of six children under 12 months three died (Table V). The problem of operating on small babies is that of using extracorporeal

TABLE V
AGE AT MUSTARD'S OPERATION AND HOSPITAL DEATHS

Age at Operation (mth)	No. of Patients	Hospital Deaths
0-12	6	3
13-24	12	2
25-36	6	0
37-48	7	2
49-60	3	1
61+	3	1
	37	10

circulation. Dillard and others (1969) have operated on children less than 6 months old using deep hypothermia and surface cooling which allowed total circulatory arrest for periods of up to 60 minutes without extracorporeal circulation. Seven infants were described, their ages ranging from 3 days to 11 months and their weights 3.0 to 6.9 kg. Four survivors have been followed up for between 12 and 30 months. Of the three who died, two had severe pulmonary hypertension, and the third a hypoplastic left ventricle. Breckenridge *et al.* (1972) reported a successful operation using cardiopulmonary bypass in an infant 7 weeks old (4.5 kg) and showed that the results of elective operation in infants under 12 months of age compared favourably with those of their whole series.

Operation may be needed before the child reaches the optimal weight, especially as delay may not result in significant additional growth of these children (Waldhausen *et al.*, 1970).

The mortality of Mustard's operation in our hands has not been as low as was originally expected (Parsons *et al.*, 1971). There were 10 hospital deaths, a mortality of 27%; of these

seven (18.9%) had uncomplicated TGA. Mustard (1964) reported five survivors out of seven. In a review of a series of 65 patients operated upon at the Mayo Clinic between 1964 and 1967 there were 25 hospital deaths, 35% (Danielson *et al.*, 1971). The majority of these cases were complicated and there were only three deaths out of 25 uncomplicated cases. Aberdeen (1971) reported 10 deaths out of 78 (13%) uncomplicated cases from the Hospital for Sick Children, London, and in a recent series of 200 from the same hospital there was an overall hospital mortality of 18% and of 11% in the uncomplicated cases (Breckenridge *et al.*, 1972).

Some postoperative bleeding is not uncommon after operation in patients with cyanotic congenital heart disease (Ekert, Gilchrist, Stanton, and Hammond, 1970). Danielson and others (1971) reported six re-explorations for postoperative bleeding, which was a relatively frequent problem in their series. The report of Clarke and Barratt-Boyes (1967) emphasizes that the chest should be re-explored when the pulmonary oedema and hypotension are associated with wound bleeding.

Dysrhythmia in the immediate postoperative period is common, sometimes contributing to death but often transitory and of no consequence. Mustard (1964) mentioned transitory dysrhythmia, and in a further report stated that dysrhythmia disappeared four to six weeks after operation (Mustard, 1968). Six of the 12 patients reported by Waldhausen and others (1970) were in nodal rhythm at the end of the operation. Only one needed transient cardiac pacing and all left hospital in sinus rhythm. Zuberbuhler and Bauersfeld (1967) reported two cases of unusual dysrhythmia; one had a variable degree of sinoatrial block and the other had two levels of block in the atrioventricular node. These dysrhythmias were attributed to the suture line interfering with the sino-atrial or atrioventricular nodes. Both patients had excellent results from the operation even though the dysrhythmia persisted. By modification of the operative technique, including placing sutures away from the atrioventricular node and the bundle of His, Breckenridge *et al.* (1972) demonstrated a significant reduction in the incidence of atrial flutter and atrioventricular dissociation, a significant cause of morbidity and mortality in their series. Complete atrioventricular dissociation occurred in five cases reported by Danielson *et al.* (1971), all of whom had repair of an additional ventricular septal defect. They all died. One of our six cases of atrioventricular block had a ventricular septal defect while the

others had uncomplicated transposition of the great arteries.

Parsons *et al.* (1971) reported 32 deaths out of 65 infants after palliative procedures and expressed the hope that earlier Mustard's operation might improve the prognosis. Our experience seems to support this.

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