Results over a Two-year Period in Complete Transposition of the Great Arteries after Mustard's Procedure

HITOSHI SHIROTANI

Postoperative echocardiographic, hemodynamic and cineangiographic findings were obtained on 27 cases of complete transposition of the great arteries (TGA) which had undergone Mustard's procedure over two years ago.

A majority of patients with TGA have abnormal right ventricular function as indicated by an increased right ventricular end-diastolic volume (RVEDV) and a low ejection fraction (RVEF). Despite this depressed RVEF, normal or increased RV output is maintained through an increased RVEDV.

The postoperative right ventricular end-diastolic dimension (RVDd) averaged $275 \pm 70\%$ of normal and the left ventricular end-diastolic dimension (LVDd) averaged $75 \pm 11\%$ of normal. The postoperative ratio of RPEP/RVET was greater (0.44 ± 0.06) than normal value (0.34 ± 0.06) for systemic circuit, whereas the postoperative ratio of LPEP/LVET was similar (0.23 ± 0.06) to normal value (0.22 ± 0.05) for pulmonary circuit. The ventricular cavity dimensions and ratios of PEP/ET were appropriate for the physiologic role of the ventricles rather than their morphologic identity.

A variety of valve motion abnormalities were noted; these included diastolic flutter of the mitral valve in 94% of patients, prolapse in 50% and systolic anterior motion (SAM) in 50%, while diastolic flutter of the tricuspid valve in 75% of patients, prolapse in 100% and SAM in 19%. Systolic flutter of the semilunar valve was found in half patients and paradoxical septal motion was found in all patients.

Atrioventricular valve motion abnormalities presumably caused by the decrease of atrial volume or compliance or the increase of RV pressure were observed at high rate but at least until 8 years after Mustard's procedure, the function of those valves had been satisfactory.

Echocardiographic dimensions or systolic time intervals (STI) were considered to be useful not only for the evaluation of LV performance but also for the evaluation of RV performance or dynamic status of pulmonary circulation after Mustard's operation for TGA.

The finding of a great number of echocardiographic abnormalities in TGA suggests that these echocardiographic features are to be expected after Mustard's procedure.

Key Words:

Transposition of the great arteries
Mustard's procedure
Echocardiography
Right ventricular volume
Cavity diameter
Systolic time interval
Systolic anterior motion
Valve flutter

THE following 3 problems have been experienced during the remote period after Mustard's procedure for complete transposition of the great arteries (TGA).

- (1) dysrhythmia
- (2) systemic and/or pulmonary venous obstruction

Department of Cardiovascular Surgery, Kinki University School of Medicine, Osaka, Japan

TABLE I MUSTARD PROCEDURE FOR COMPLETE TRANSPOSITION OF THE GREAT ARTERIES

	TGA I	TGA II
Total	28	31
30 days survivors	22	17
Long time survivors (> 2yrs)	15	12
Age (mean)	3y2m - 8y9m (5y4m)	4y4m - 12y11m (7y 4m)
Follow-up time (mean)	2y2m - 8y2m (4y5m)	3y9m - 6y 4m (4y10m)

- (3) dysfunction of tricuspid valve and right ventricle due to the use of right ventricle (RV) as systemic pumping chamber

In the present study, the aforesaid 3 problems especially the problem No.3 has been considered in reference to the electrocardiographic echocardiographic hemodynamic and cineangiographic findings.

MATERIALS AND METHODS

The cases studied this time were 27 cases of successful Mustard's procedure including 15 cases of TGA with intact ventricular septum (Group I) and 12 cases of TGA with large VSD

(Group II) which passed 2 years or more after operation.

The age of patients in Group I ranged from 3 - 2/12 years to 8 - 9/12 years (mean 5 - 4/12 years) and in Group II from 4 - 4/12 years to 12 - 11/12 years (mean 7 - 4/12 years).

The period of postoperative follow-up was 2 - 2/12 years to 8 - 2/12 years (mean 4 - 5/12 years) for Group I and 3 - 9/12 years to 6 - 4/12 years (mean 4 - 10/12 years) for Group II (Table I).

Echocardiography was applied to 8 cases each of Group I and Group II, in total 16 cases. Echocardiograms were performed with a Toshiba

Fig.1. Echocardiogram showing method by which right ventricular systolic time intervals (STI) are measured from aortic and tricuspid valve motion in TGA. Sagittal cross-sectional echocardiograms (left figure) demonstrates two different M-mode echoes with different beam directions passing through the aortic and tricuspid valves (1 and 2). The lines drawn indicate the technique used to measure STI. PEP = pre-ejection period; ICT = isovolumic contraction time; RVET = right ventricular ejection time; AoV = aortic valve; TV = tricuspid valve; RV = right ventricle; AO = aortic root; SVA = systemic venous atrium.

Fig. 2. The ratio of the pulmonary to systemic systolic pressure (P_p/P_s) before and after Mustard's procedure.

Model SSH-11A sonolayergraph, coupled to a Honeywell Model 1219 strip chart recorder.

Right ventricular volumes were calculated from biplane cineangiograms using Simpsons rule method¹ and right ventricular end-diastolic volume (RVEDV) were compared to the normal ranges established by Nakazawa et al.² The diastolic dimensions of the right and left ventricles were compared to the normal ranges established by Epstein et al.³

Systolic time intervals (STI) for both ventricles were obtained from the ECG and from semilunar valve leaflet opening and closing⁴ as shown in Figure 1. The ratio of the pre-ejection period to the ejection time (PEP/ET) for each ventricle was calculated using the average values from 3 cardiac cycles.

RESULTS

Electrocardiographic Findings

Most cases recovered sinus rhythm in two weeks after operation or at the latest in two months. The cases which accompanied some kind of arrhythmia counted only three and even in those three cases, the anomaly was temporary junctional rhythm or atrial premature beat.

One case of temporary junctional rhythm, however, died suddenly presumably due to

Fig. 3. Mean $\pm$ SD for cineangiographic right ventricular end-diastolic volume (RVEDV) (expressed as % of the predicted normal) in TGA patients before and after Mustard's procedure. Normal values are shown by the shaded area.

Fig. 4. The right ventricular ejection fraction (RVEF) in TGA patients before and after Mustard's procedure. The shaded area represents the mean $\pm$ 2SD for normal ranges.

arrhythmia one year after the operation. Therefore the follow-up of dysrhythmia seems to be quite important.

Complete right bundle branch block was observed in 25% of the cases in Group II. They were all the cases of closure of VSD by right ventriculotomy.

Both Group I and II indicated remarkable right ventricular hypertrophy and $R_{V_1} + S_{V_6}$ was 4.3 mV in the average.

Hemodynamic Findings

Figure 2 shows the transition of the ratio of pulmonary to systemic systolic pressure (P_p/P_s) before and after Mustard's procedure. In Group

Fig.5. Observed right ventricular end-diastolic dimension (RVDd) and left ventricular end-diastolic dimension (LVDd) after Mustard's procedure, and corresponding predicted normal values plotted against body surface area (BSA). The solid lines and broken lines indicate the normal values of LVDd and RVDd, respectively.

Fig.6. Mean $\pm$ SD for RVDd and LVDd (% of normal) in TGA patients after Mustard's procedure.

I, two cases of complication of PDA and one other case indicated P_p/P_s of over 0.8 before operation but pulmonary artery pressure decreased remarkably after operation and mean value of P_p/P_s decreased from 0.64 before operation to 0.29 after operation.

In Group II, mean P_p/P_s decreased from 0.98 before operation to 0.45 in 2 – 3 months after operation. In the remote period of over 2 years after operation, it decreased to a mean of 0.37.

The mean cardiothoracic ratio in the remote period of over 2 years of operation was 0.55 for Group I and 0.53 for Group II.

The systemic and pulmonary venous obstruction following Mustard's procedure has been reported previously^{5,6,7}. Pulmonary venous obstruction was not observed in this study. But only one case developed severe obstruction of SVC and IVC and he had successful surgical relief 4 years after Mustard's operation.

Venous obstruction in the earlier postoperative period may be avoided by selecting appropriate operative technique such as excision of atrial septum and devising appropriate size, shape and method of suture of atrial baffle⁸. Venous obstruction in remote period may be caused by formation of cicatrix and its contraction which are possible due to infection of the baffle and atrial wall^{8,9}. Attention must be paid to the prevention of postoperative infection.

Cineangiographic Findings

(1) Right Ventricular End-diastolic Volume (RVEDV)

RVEDVs were larger than normal in both Group I and II, and tended to decrease postoperatively as shown in Figure 3. The average value for RVEDV was from $183 \pm 50\%$ to $142 \pm 27\%$.

Fig. 7. The ratios of the pre-ejection period to ventricular ejection time (PEP/ET) for systemic and pulmonary circuits in TGA patients after Mustard's procedure. The broken line represents those in patients with normal heart (left figure). The mean pulmonary artery pressure (MPAP) is plotted against LPEP/LVET in postoperative TGA patients (right figure).

of normal in Group I and from $165 \pm 34\%$ to $154 \pm 23\%$ of normal following Mustard's operation in Group II.

(2) Right Ventricular Ejection Fraction (RVEF)

Figure 4 illustrates the RVEF for both groups. The preoperative RVEF averaged 0.55 ± 0.14 in Group I and 0.63 ± 0.08 in Group II. The average value in Group II markedly decreased following Mustard's operation while a small decrease was found in Group I.

Echocardiographic Findings

(1) Right Ventricular End-diastolic Dimensions

(RVDd) and left Ventricular End-diastolic Dimensions (LVDd)

RVDd were strikingly larger than normal before and after operation in both groups, and approached the values of the normal left ventricle (LV). In contrast, LVDd were significantly reduced postoperatively (Fig. 5).

Figure 6 illustrates RVDd and LVDd for both groups. The postoperative RVDd averaged $275 \pm 70\%$ of normal and the postoperative LVDd averaged $75 \pm 11\%$ of normal.

(2) Systolic Time Intervals (STI)

The ratio of the pre-ejection period to the

TABLE II ECHOCARDIOGRAPHIC ABNORMALITIES IN 16 PATIENTS FOLLOWING MUSTARD PROCEDURE

		TGA I	TGA II	Total
Diastolic flutter	Mit.	100%	88%	94%
	Tri.	88	63	75
Prolapse	Mit.	25	75	50
	Tri.	100	100	100
SAM*	Mit.	63	38	50
	Tri.	25	13	19
Systolic flutter	Ao.	63	43	53
	Pul.	29	86	57

*systolic anterior motion

ejection time (PEP/ET) for each ventricle is shown in Figure 7. The PEP/ET was greater in the systemic than in the pulmonary circuit. RPEP/RVET was 0.44 ± 0.05 and LPEP/LVET was 0.23 ± 0.06 in Group I. RPEP/RVET was 0.45 ± 0.06 and LPEP/LVET was 0.23 ± 0.07 in Group II. The postoperative RPEP/RVET was greater than normal value (0.34 ± 0.06) for systemic circuit, whereas the postoperative LPEP/LVET was similar to normal value (0.22 ± 0.05) for pulmonary circuit.

The postoperative LPEP/LVET was correlated with the mean pulmonary artery pressure (MPAP) at cardiac catheterization. The correlation coefficients relating LPEP/LVET to MPAP was 0.80. (3) Atrioventricular and Semilunar Valve Motion, and Ventricular Septal Motion

The atrioventricular and semilunar valve echoes showed several abnormalities (Table II). Diastolic flutter of mitral valve was present in 15 of 16 patients, and 12 had tricuspid flutter. Prolapse of tricuspid valve was present in all, while 8 had mitral prolapse. Systolic anterior motion (SAM) of mitral valve was observed in 8, while 3 had SAM of tricuspid valve. Semilunar valve flutter in systole was present in a little over one half of the cases.

Paradoxical septal motion was found in all and was similar to that described in patients with left bundle branch block.¹⁰

DISCUSSION

It is not known how long RV can continue to function as a systemic ventricle, or whether long-term baffle problems will develop. In view of these problems, the TGA patients even after Mustard's operation will require continuous monitoring throughout life.

The elevated RVEDV and depressed RVEF were present in the majority of patients with TGA. The depressed RVEF is unlikely to be due to abnormal loading conditions (an increased afterload or a decreased preload) and is probably due to depressed RV function. Despite this depressed RV function, normal or in some cases increased RV output is maintained through the Starling mechanism as a result of an increased RVEDV. These findings are in agreement with similar data published by Jarmakani et al.¹¹ and Graham et al.¹²

Most patients had increased RVDd, reflecting RV dilatation found at angiography. The altered dimension is reflected by an increase in antero-posterior dimension. The small LVDd may

reflect the lower afterload of the pulmonary ventricle after Mustard's operation.

The effects of the natural reversal of systemic and pulmonary circuits on RV and LV was observed in 16 patients with TGA. There was a prolonged RPEP and shortened RVET, producing a high ratio of RPEP/RVET. This may have been the result of increased afterload of systemic pressure and may also be a result of myocardial dysfunction. In two out of three patients with RPEP/RVET of 0.5 or greater, angiographic evidence of myocardial dysfunction was observed and they required the management with such drugs as digitalis and furosemide. Such high ratio of RPEP/RVET appeared to indicate that RV is poorly adapted to assume the function of a systolic ventricle.

Hirschfeld et al.¹³ have demonstrated that LPEP/LVET in TGA can be used to estimate pulmonary artery pressure and resistance. The present study demonstrated that the postoperative LPEP/LVET was directly related to MPAP and marked elevation of the ratio indicated the presence of pulmonary hypertension.

A large number of valve motion abnormalities were found in patients who have undergone Mustard's procedure; these included diastolic flutter and SAM of the atrioventricular valves, systolic flutter of the semilunar valves and paradoxical septal motion. These findings have been reported in several report series.^{14,15,16} Atrioventricular valve fluttering has been attributed to the relatively restrictive atrial tunnel, resulting in turbulent blood flow across these valves.¹⁶ SAM of the mitral valve and systolic flutter of the pulmonary valve may be due largely to bulging of the ventricular septum into LV cavity with dynamic LV outflow narrowing. The high velocity flow through the narrowed LV outflow tract may have caused further anterior displacement of the mitral valve in systole by a Bernoulli effect,^{14,15} mimicing idiopathic hypertrophic subaortic stenosis.^{17,18} The turbulence beyond the area of the outflow narrowing probably produces pulmonary valve flutter.

Tricuspid incompetence has been reported in postoperative TGA patients¹⁹ and is another factor which might contribute to abnormal RV function. Although prolapse of the tricuspid valve was found in all patients, 3 of 27 patients in the present study had significant tricuspid incompetence as judged both by cineangiography and by physical examinations.

Since serial echocardiograms can be per-

formed safely and rapidly on an out-patient basis, it is particularly useful for longitudinal studies of individual patients following Mustard's procedure.

REFERENCES

1. GRAHAM TPJr, JARMAKANI JM, ATWOOD GF, CANENT RVJr: Right ventricular volume determinations in children, normal values and observations with volume or pressure overload. *Circulation* 47: 144, 1973
2. NAKAZAWA M, MARKS RA, ISABEL-JONES J, JARMAKANI JM: Right and left ventricular volume characteristics in children with pulmonary stenosis and intact ventricular septum. *Circulation* 53: 884, 1976
3. EPSTEIN ML, GOLDBERG SL, ALLAN HD, KONECKE L, WOOD J: Great vessel, cardiac chamber and wall growth patterns in normal children. *Circulation* 51: 1124, 1975
4. HIRSCHFELD S, MEYER R, SCHWARTZ DC, KORFHAGEN J, KAPLAN S: Measurement of right and left ventricular systolic time intervals by echocardiography. *Circulation* 51: 304, 1975
5. STARK J, TYNAN M, ASHCRAFT KW, ABERDEEN E, WATERSTON DJ: Obstruction of pulmonary veins and superior vena cava after Mustard operation for transposition of the great arteries. *Circulation* 45 & 46 (Suppl I): I-116, 1972
6. CHAMPSAUR GL, SOKOL DM, TRUSLER GA, MUSTARD WT: Repair of transposition of the great arteries in 123 pediatric patients. Early and long-term results. *Circulation* 47: 1032, 1973
7. CLARKSON PM, NEUTZE JM, BARRATT-BOYES BG, BRANDT PWT: Late postoperative hemodynamic results and cineangiographic findings after Mustard atrial baffle repair for transposition of the great arteries. *Circulation* 53: 525, 1976
8. SHIROTANI H, ANDO F: Postoperative problems and its countermeasures following Mustard operation for complete transposition of the great arteries with ventricular septal defect. *Operation* 30: 825, 1976 (in Japanese)
9. MAZZEI FA, MULDER DG: Superior vena cava syndrome following complete correction (Mustard repair) of transposition of the great vessels. *Ann Thorac Surg* 11: 243, 1971
10. ABBASI AS, EBER LM, MacALPIN RM, KATTUS AA: Paradoxical motion of interventricular septum in left bundle branch block. *Circulation* 49: 423, 1974
11. JARMAKANI JM, CANENT RVJr: Preoperative and postoperative right ventricular function in children with transposition of the great vessels. *Circulation* 49 & 50 (Supple II): II-39, 1974
12. GRAHAM TPJr, ATWOOD GF, BOUCEK RJr, BOERTH RC, NELSON JH: Right heart volume characteristics in transposition of the great arteries. *Circulation* 51: 881, 1975
13. HIRSCHFELD MD, MEYER R, SCHWARTZ DC, KORFHAGEN J, KAPLAN S: The echocardiographic assessment of pulmonary artery pressure and pulmonary vascular resistance. *Circulation* 52: 642, 1975
14. PARK SC, NECHES WH, ZUBERBUHLER JR, MATTHEWS RA, LENOX CC, FRICKER FJ: Echocardiographic and hemodynamic correlation in transposition of the great arteries. *Circulation* 57: 291, 1978
15. SILVERMAN NH, PAYOT M, STANGER P, RUDOLPH AM: The echocardiographic profile of patients after Mustard's operation. *Circulation* 58: 1083, 1978
16. HANGER DJ, TAJIK AJ, RITTER DG: Fluttering of atrioventricular valves in patients with d-transposition of the great arteries after Mustard operation an echographic observation. *Myo Clin Proc* 50: 69, 1975
17. KING JF, DeMARIA AN, MILLER RR, HILLIARD GK, ZELIS R, MASON DT: Markedly abnormal mitral valve motion without simultaneous intraventricular pressure gradient due to uneven mitral-septal contact in idiopathic hypertrophic subaortic stenosis. *Am J Cardiol* 34: 360, 1974
18. HENRY WL, CLARK CE, GRIFFITH JM, EPSTEIN SE: Mechanism of left ventricular outflow obstruction in patients with obstructive asymmetric septal hypertrophy (Idiopathic hypertrophic subaortic stenosis). *Am J Cardiol* 35: 337, 1975
19. TYNAN M, ABERDEEN E, STARK J: Tricuspid incompetence after the Mustard operation for transposition of the great arteries. *Circulation* 48 (Suppl I): I-111, 1972