The Echocardiographic Profile of Patients After Mustard’s Operation

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SUMMARY In order to establish an echocardiographic profile of patients with simple transposition after Mustard’s operation, we examined the M-mode records of 10 patients who were found to be free of significant abnormalities at follow-up cardiac catheterization. When compared with established normals, right ventricular wall thicknesses and cavity dimensions were increased, while left ventricular wall thicknesses and cavity dimensions fell below the mean. The wall thicknesses, cavity dimensions and ratios of right ventricular prejection period/ejection time and left ventricular prejection period/ejection time were appropriate for the physiologic role of the ventricles rather than their morphologic identity.

In each patient, a portion of the intra-atrial baffle was identified behind the pulmonary root. There was variation in baffle position and baffle mobility within the group, as well as in individual echograms. A variety of valve motion abnormalities were noted; these included diastolic flutter of the atroventricular valves in all 10 patients and systolic anterior motion of the mitral valve in six patients. Paradoxical septal motion was found in nine patients. Although only minimal or no left ventricular outflow gradients were found at catheterization, nine patients had narrowing of the left ventricular outflow tract, 10 had systolic flutter of the pulmonary valve and eight had early partial closure of the pulmonary valve.

The finding of a large number of echocardiographic abnormalities in a group of patients with good hemodynamic results suggests that these echocardiographic features are to be expected after Mustard’s operation. Furthermore, the reversal of the physiologic role of the ventricles must be considered when interpreting the echocardiographic dimensions and systolic time intervals.

PATIENTS WHO HAVE UNDERGONE Mustard’s operation have been found to have a wide variety of anatomic, hemodynamic and echocardiographic abnormalities, and each of these may change over time.1-11 Evaluation of such changes by serial cardiac catheterization is associated not only with risk but also with cumulative radiation dosage and technical problems related to peripheral vessel reentry. Although echocardiography provides more limited anatomic and physiologic information than cardiac catheterization, such noninvasive studies augment clinical evaluation in deciding an appropriate time for recatheterization. Because the cardiac anatomy and physiology of patients who have undergone Mustard’s operation is quite different from that of normal individuals, conventional echocardiographic standards may not be applicable to these patients. Although several echocardiographic abnormalities have been reported in post-Mustard patients12-16 it is not clear which of these abnormalities are indicative of a significant hemodynamic disturbance. Therefore, we have attempted to establish a qualitative and quantitative echocardiographic profile of patients with simple transposition who underwent Mustard’s operation and who, at postoperative cardiac catheterization, were found to have a good hemodynamic result.

Methods

Ten post-Mustard patients with good hemodynamic results shown by cardiac catheterization, who also had M-mode echocardiograms, were studied.
Only patients with simple transposition (that is, with intact ventricular septum and without significant pulmonary outflow obstruction or patent ductus arteriosus) were included in the study. All 10 had pericardium used to fashion the intra-atrial baffle. Their ages at the time of repair ranged from 1½ weeks–4 years. Cardiac catheterization was performed one month to six and one-half years after Mustard’s operation, and an echocardiogram was obtained within 24 hours of catheterization.

Seven patients had two or more postoperative echocardiographic studies. Serial studies have been obtained in these patients for up to two and one-half years. All echocardiograms were obtained with a SmithKline 20A Ultrasoundoscope interfaced with a strip chart recorder. Transducers were selected according to individual patient size. Tracings were recorded at paper speeds of 50 mm/sec, except for systolic time intervals, which were recorded at 100 mm/sec. When we recorded time intervals, the time lines were generated every 500 msec, and paper speed was calibrated at 100 mm/sec. The accuracy of the time line generator was checked electronically and varied less than 1%. Measurements were made with a metric ruler and were accurate to within 5 msec.

In each patient, the thickness of the left ventricular posterior wall, right ventricular anterior wall and ventricular septum were measured at end-diastole. The diastolic dimensions of the left and right ventricles, aortic root and pulmonary arterial root at the level of their respective valve leaflets, as well as the atrioventricular valve D to E excursions, were measured. For each variable, three separate complexes were measured and their average values were expressed to the nearest 0.5 mm. Dimensions were compared to the normal ranges for children established in our laboratory. All four valves were found in the expected positions for d-transposition. The echoes arising from the intra-atrial baffle were identified in each patient.

Systolic time intervals for both ventricles were obtained from the ECG and from semilunar valve leaflet opening and closing. Using the average values from three cardiac cycles, we calculated the ratio of the pre-ejection period to the ejection period for each ventricle.

Qualitative assessments were made of ventricular septal motion, baffle position and left ventricular outflow tract narrowing. The ventricular septal motion was determined to be normal or paradoxical just below or at the level of the mitral valve leaflets. A portion of the baffle behind the pulmonary artery was identified and its position within the atrium at end-diastole was displayed in each case, as was the vigor of baffle motion.

Left ventricular outflow tract narrowing was assessed by measuring from the mitral C point to the ventricular septum during an echocardiographic sweep from the left ventricle to the pulmonary artery root. The dimension of the narrowest portion of the outflow tract was expressed as a percentage of the pulmonary arterial root diastolic dimension.

Descriptions of abnormalities of semilunar and atrioventricular valve motion were also included.

In order to define the echocardiographic profile for simple transposition post-Mustard with good hemodynamic results, several patients were excluded from this study. Those with iliofemoral venous obstruction that precluded recatheterization from the groin (six patients) were not recatheterized unless there was clinical evidence of significant abnormality. In addition, those with significant hemodynamic disturbances at recatheterization were also excluded from this study. The latter included pulmonary vascular disease (one patient), mild pulmonary hypertension (two patients), tricuspid insufficiency (one patient), brady-arrhythmias (two patients) and subpulmonary stenosis greater than 25 mm Hg (two patients). Because almost all post-Mustard patients have mild subpulmonary outflow gradients, we included patients with systolic gradients of 25 mm Hg or less in this study. Minute baffle leaks detectable only by cineangiography and mild superior vena caval obstructions were considered to be insignificant from a hemodynamic standpoint and therefore were not reasons for exclusion from the study. Superior vena caval obstruction was considered mild if there was angiographic patency of the superior limb of the baffle, no runoff via the azygos vein, and a gradient of less than 3 mm Hg.

Results

Cardiac Catheterization Data

The pertinent data obtained at cardiac catheterization for each of the 10 patients is summarized in table 1. These data included right ventricular, left ventricular and pulmonary arterial pressures, left ventricular outflow gradients and the presence or absence of superior vena caval obstruction. In addition, we examined the patients’ ventricular angiograms; each showed a large right ventricle with reduced ejection fraction, findings that have been reported in patients after Mustard’s operation.

Echocardiographic Data

The quantitative echocardiographic data are tabulated in table 2 and qualitative data are summarized in table 3. Representative echocardiograms are shown in figures 1–3.

In eight of 10 patients, the right ventricular wall thickness was above the 95% tolerance limits for normal individuals. Right ventricular wall thickness in all 10 patients, however, fell within the range of the normal for left ventricles. In all 10 patients, left ventricular wall thickness was within the 95% tolerance limits for normal left ventricles; however, the values clustered in the low range of normal. The physiologic role of the ventricles is reversed in transposition (i.e., the right ventricle faces the systemic resistance, the left ventricle a pulmonary resistance). Consequently, ventricular wall thicknesses were also compared with normals for the contralateral ventricles. When the left
ventricular wall thickness was compared to right ventricular normals, the left ventricular wall thickness was either high normal or increased. Septal thickness was normal in all 10 patients; however, the values clustered in the high normal range.

Cavity Dimensions

Right ventricular cavity dimensions were high normal in two patients and above the 95% tolerance limits in eight patients (fig. 4). In contrast, all 10 right ventricular cavity dimensions were in the normal range for a normal left ventricle. Left ventricular cavity dimensions were in the lower normal range in seven patients and below normal in three patients. Left ventricular cavity dimensions were either high normal or increased when compared with right ventricular normals.

Root Dimensions

All 10 patients had pulmonary roots of normal size. Aortic roots were in the upper range of normal in five patients and above the 95% tolerance limits in five patients. In nine of 10 cases the aortic root was larger than the pulmonary root. This may be due to oblique transection of the aortic root by the echo beam.

Ventricular Septal Motion

There was an abnormality of ventricular septal motion in nine of 10 patients. In each of the nine cases, the septal motion was similar to that described in patients with left bundle branch block.21, 22 At the end of the electrocardiographic QRS complex the septum moved abruptly posteriorly; at the time of mitral closure the septal motion became flat and remained so for the remainder of systole (figs. 1 and 5).

<table>
<thead>
<tr>
<th>Table 1. Post-Mustard Catheterization</th>
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<tr>
<td>Patient no.</td>
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<td>9</td>
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Numbers with bars indicate mean values.

Abbreviations: RV = right ventricle; LV = left ventricle; PA = pulmonary artery; LVOG = peak systolic left ventricular outflow gradient; IVC = inferior vena cava; SVC = superior vena cava.

<table>
<thead>
<tr>
<th>Table 2. Echocardiographic Dimensions and Time Indices</th>
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<td>9</td>
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<td>10</td>
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</table>

The patient sequence is the same as table 1.

Abbreviations: RV = right ventricle; LV = left ventricle; Ao = aorta; PA = pulmonary artery; PEP/ET = preejection period to ejection time ratios for right ventricle (RV) and left ventricle (LV).
Table 3. Qualitative Echocardiographic Abnormalities in 10 Patients who Underwent Mustard’s Operation

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Ventricular septal motion</th>
<th>Mitral SAM</th>
<th>Ratio of LV outflow to pulmonary Artery dimension</th>
<th>Pulmonary valve flutter</th>
<th>Diastolic flutter</th>
<th>Baffle</th>
<th>Amplitude of motion</th>
<th>Position in atrium</th>
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<td>1</td>
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<td>0</td>
<td>0.84</td>
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<td>+</td>
<td>+</td>
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<td>Mild</td>
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<tr>
<td>2</td>
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<td>0</td>
<td>0.75</td>
<td>+</td>
<td>+</td>
<td>+</td>
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<tr>
<td>3</td>
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<td>Mild</td>
<td>0.75</td>
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<td>+</td>
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<td>0</td>
<td>Midposterior</td>
</tr>
<tr>
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<td>0.80</td>
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<td>+</td>
<td>+</td>
<td>0</td>
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</tr>
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<td>Abnormal</td>
<td>Moderate</td>
<td>0.75</td>
<td>+</td>
<td>+</td>
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<tr>
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<td>+</td>
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<td>Mid</td>
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<td>+</td>
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<td>0.83</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>Mild</td>
</tr>
<tr>
<td>9</td>
<td>Abnormal</td>
<td>Mild</td>
<td>1.00</td>
<td>+</td>
<td>0</td>
<td>+</td>
<td>0</td>
<td>Flat</td>
</tr>
<tr>
<td>10</td>
<td>Abnormal</td>
<td>Mild</td>
<td>0.83</td>
<td>+</td>
<td>0</td>
<td>0</td>
<td>+</td>
<td>Midposterior</td>
</tr>
</tbody>
</table>

# Abnormal 9/10 6/10 9/10 10/10 8/10 9/10 4/10 6/10

% Abnormal 90% 60% 90% 100% 80% 90% 40% 60%

Abbreviations: SAM = systolic anterior motion; 0 = absent; + = present.

Atrioventricular Valve Motion

The atrioventricular valve echoes showed several abnormalities, including diastolic flutter. Mitral valve diastolic flutter was present in nine patients (fig. 1). Although both leaflets showed flutter, it was generally more marked in the anterior leaflet. Tricuspid anterior leaflet flutter was present in four patients and three of these patients also had posterior leaflet flutter.

The amplitude of the mitral leaflet D-E excursion was within the normal range in all 10 patients; however, in each case the amplitude was below the mean for normal (fig. 4). Tricuspid D-E excursion clustered in the upper range of normal. Systolic anterior motion of the mitral valve was present in seven of 12 patients (table 3). The presence or absence of this finding was not related to subpulmonary gradient.

Subpulmonary Narrowing

We assessed left ventricular outflow tract narrowing echocardiographically, using the criteria of Aziz et al. The ratio of the narrowest left ventricular outflow measurement to pulmonary arterial root dimension was between 0.75 and 1 (table 3), values that would not be expected to have significant associated outflow gradients. In each of these patients the left ventricular outflow tract appeared narrow in the lateral projection of left ventricular cineangiogram (fig. 6); however, there appeared to be no relationship between outflow gradient and echocardiographic assessment of outflow tract narrowing. It is possible that echocardiography assesses the narrowest diameter of an elliptical outflow and therefore may not be representative of total cross-sectional area. The left ventricular outflow tract/pulmonary artery ratio does not appear to be a reliable indicator of mild or no obstruction at catheterization.

Intra-atrial Baffle

In all 10 patients, dense echoes from the intra-atrial baffle were identified within the atrium when the echo beam passed through the pulmonary root (fig. 7). Echoes from the baffle also were noted posterior to the tricuspid valve in four patients and posterior to the aortic root in three patients. When a sweep was made from the pulmonary valve to the mitral valve, the baffle echo fused with the echo of the posterolateral portion of the mitral annulus. Although other small portions of the intra-atrial baffle occasionally were detected behind the tricuspid valve or behind the aorta, a comprehensive view of the superior and inferior limbs of the baffle could not be obtained in any patient.

The upper portion of baffle is seen behind the pulmonary root. The atrium directly anterior to the baffle is the upper limb of the systemic venous atrium and the posterior is the portion of pulmonary venous atrium that lies adjacent to the pulmonary veins. The anteroposterior position of this portion of baffle is indicated in table 3. The position of the baffle did not appear to be related to superior vena caval obstruction.

The amplitude of the baffle motion varied considerably, not only among the patients but also within individual echocardiograms, depending on transducer position.

Systolic Time Intervals

The ratio of the preejection period to the ejection time (PEP/ET) for each ventricle is shown in figure 8 and table 2. The ratio of the preejection period to the
**Figure 1.** Representative tracing from patient 10 after Mustard's operation, showing the plane used for measuring ventricular cavity dimensions and wall thicknesses. Abbreviations: RVW = right ventricular wall; RVC = right ventricular cavity; Sept = ventricular septum; LVC = left ventricular cavity; LVW = left ventricular posterior wall. Arrows indicate tricuspid valve leaflets within RVC and mitral valve within the LVC. Flutter on the atroioventricular valve leaflets is noted.

**Figure 2.** Aortic root (Ao) echogram from patient 6, showing the aortic leaflets. The lines drawn indicate the technique used to measure the systolic time intervals for the right ventricle, pre-ejection period (PEP) and ejection time (RVET). The technique for measuring these intervals using phonocardiogram (PCG) and carotid pulse (CAR) are shown for comparison.
Ventricular ejection time was greater in the systemic than in the pulmonary circuit in all 10 patients. The mean values for the systemic and pulmonary PEP/ET ratios were similar to those reported for d-transposition before Mustard's operation. Right ventricular PEP/right ventricular ET was 0.35 ± 0.08 and left ventricular PEP/left ventricular ET was 0.21 ± 0.8 (fig. 8). The ratios obtained from the systemic circuit were always greater than those of the pulmonary circuit ($P < 0.01$, paired $t$ test) and were similar to normal values for systemic and pulmonary circuits, respectively. 19, 24, 25

Six of these patients have been followed with serial echocardiograms after Mustard's operation. The echocardiographic abnormalities were constant on repeat examinations obtained over a period of three months to two and one-half years. Abnormalities of valve motion have persisted, as have baffle motion characteristics, baffle position and left ventricular outflow tract narrowing. Additionally, the echocardiographic dimensions showed appropriate changes with patient growth.
Discussion
A large number of echocardiographic abnormalities have been found in patients who have undergone Mustard's operation; however, no systematic correlation has previously been made with hemodynamic status. In order to define which of these echo abnormalities are intrinsic to the altered morphology and rerouted circulations, we have examined the echo-

**Figure 4.** Graph indicating normal values for body surface area (BSA) in square meters (m²). The continuous lines indicate the 5th and 95th % tolerance lines and the mean (center). Data points are those of the patients with aortopulmonary transposition for their specific body surface area. RVAWD = right ventricular anterior wall at end-diastole; RVDD = right ventricular cavity end-diastolic dimension; Sept D = septal diastolic dimension; MVDE = mitral D-E excursion; LVEDD = left ventricular end-diastolic dimension; LVPWD = left ventricular posterior wall end-diastolic dimension; TVDE = tricuspid D-E excursion; AOD and PAD = aortic and pulmonary artery root diastolic dimension.

**Figure 5.** Echogram from a patient (no. 10) with aortopulmonary transposition showing systolic anterior motion of the mitral valve (SAM). RVW = right ventricular anterior wall; RVC = right ventricular cavity; Sept = ventricular septum showing paradoxical septal motion; LVC = left ventricular cavity; LVW = left ventricular wall.
cardiograms of 10 patients who have been shown to have good surgical results both by clinical assessment and cardiac catheterization. The echocardiographic profile of these 10 patients represents the typical findings in the postoperative Mustard patient who currently has a satisfactory postoperative state.

Included in this group are patients with obstruction of the superior limb of the systemic venous atrium, mild left ventricular outflow gradients and tiny baffle leaks, because almost all post-Mustard patients have one or more of these minor abnormalities. These findings have been found in several reported series.2, 3, 7, 8

Figure 6. Systolic frame of left ventricular angiogram. Posteroanterior position on the left and lateral on the right. The arrows indicate the area of subpulmonary narrowing and narrow posteroanterior ventricular dimensions related to the displacement of the ventricular septum by the enlarged right ventricle.

Figure 7. Echocardiogram from patient 5, showing the pericardial baffle. PA = pulmonary artery; PV = pulmonary valve; Sept = interventricular septum; LVW = left ventricular wall.
and the patients in our own group have been operated on by several different surgeons from four centers. The minor residuals, therefore, do not reflect the technique of a particular surgeon.

There are a number of echocardiographic features that appear to be constant after Mustard's operation. Most notable are the abnormal wall thicknesses and ventricular chamber dimensions. The increase in right ventricular wall thickness is not surprising, since the right ventricle has functioned as the systemic ventricle both before and after surgery. When one compares right ventricular wall thicknesses with left ventricular normals, it appears that the degree of thickness is appropriate for a ventricle facing systemic afterload. The observed values for left ventricular wall thicknesses were generally low for a morphologic left ventricle; all fell below the mean for normal left ventricles. The relatively thin left ventricle in patients with transposition may be related to the lower afterload faced by a pulmonic ventricle.

Angiographic volume studies of post-Mustard patients have shown increased right ventricular end-systolic and end-diastolic volumes, as well as decreased ejection fractions. Most patients had increased right ventricular dimension (fig. 4), reflecting the right ventricular dilatation found at angiography. This altered dimension is reflected by an increase in the anteroposterior dimension, as occurs in atrial septal defects and as was noted in our preliminary two-dimensional echocardiographic studies. The small left ventricular cavity dimensions may reflect the response of the morphologic left ventricle to the lower preload of the pulmonary ventricle after surgical correction. The measurements seem to reflect actual changes in size rather than the echo beam transecting different portions of the ventricle in view of the two-dimensional echocardiographic findings.

Left ventricular outflow tract narrowing both with and without systolic gradient is a frequent finding at catheterization after Mustard's operation. These patients were selected to have either no gradient or a minimal gradient. The echocardiographic findings of a narrow left ventricular outflow tract, systolic anterior motion of the mitral valve, pulmonary valve flutter

**Figure 8.** The ratios of pre-ejection period to ventricular ejection time (PEP/ET) for the left side (pulmonic) and the right side (systemic) ventricles. Lines are drawn between the two data points for each patient. Open circle and bars indicate mean and standard deviation for each group of variables.
and partial systolic closure were present alone or in combination in all of these patients. Therefore, the presence of these findings after Mustard’s operation need not indicate significant left ventricular outflow obstruction. The significance of these findings before Mustard’s operation is unclear, as there are conflicting reports.\textsuperscript{16, 23} Nanda et al.\textsuperscript{23} found a positive correlation between these findings and left ventricular outflow obstruction, while Park et al.\textsuperscript{16} found similar abnormalities in the echocardiogram of patients with transposition before and after Mustard’s operation, including those with low left ventricular systolic pressures. The latter, as well as our own findings, suggest that these abnormalities may merely reflect turbulent flow and need not be associated with a significant pressure gradient.

Although the cause of the above echocardiographic features post-Mustard is uncertain, it is possible that they are related to the posteriorly directed ventricular septal convexity encroaching on the left ventricular outflow tract (fig. 6). The mitral and pulmonary valve abnormalities may be explained by mechanisms similar to those proposed for the abnormal mitral and aortic valve movements found in idiopathic hypertrrophic subaortic stenosis.\textsuperscript{27, 28} High velocity flow through the narrowed left ventricular outflow tract produces a Bernoulli effect which in turn draws the anterior mitral leaflet toward the ventricular septum. This dynamic left ventricular outflow narrowing may transiently reduce systolic flow and cause early-systolic partial closure of the pulmonary valve. The turbulence beyond the area of the outflow narrowing probably produces pulmonary valve flutter.

Paradoxical motion of the ventricular septum was observed in nine patients. There are several theories concerning the etiology of paradoxical septal motion and at least two of these may be operative in post-Mustard patients. First, movement of the ventricular septum to the center of a common ventricular mass has been noted in patients with right ventricular enlargement from a variety of causes including those with transposition that have not undergone surgery.\textsuperscript{29, 30} Second, such septal motion has been found in patients with congenital or surgically acquired absence of the pericardium.\textsuperscript{31} Post-Mustard patients have both enlarged right ventricles and absent pericardium, a portion of the latter being used to form the intra-atrial baffle.

M-mode echocardiography provides limited visualization of the intra-atrial baffle and of the pulmonary and systemic venous atria. This is not surprising because the three-dimensional configuration of the baffle is quite complex. Due to its greater ability to define spatial interrelationships, two-dimensional echocardiography has proven more useful in visualizing these structures.\textsuperscript{26}

Considerable variation of the degree of baffle motion was noted even within a given study. This may reflect imaging of different portions of the baffle, i.e., portions close to suture lines may move less than do more central portions of the baffle. When baffle motion was vigorous, the pattern of the motion was similar to that of the atrioventricular valves with “E” and “A” points being identified (fig. 7). The latter may be the result of phasic flow within the atria, or may reflect atrial wall motion.

In this selected group of patients, the ratios of systolic time intervals for the right and left ventricles were appropriate for normal systemic and pulmonary ventricles, respectively. It appears, therefore, that in post-Mustard patients, systolic time interval ratios should be compared with standards appropriate for the physiologic role of the ventricles.

The purpose of this study was to define the post-Mustard echocardiographic norms using catheterization findings for comparison. Although it would have been interesting to compare pre- and post-Mustard echocardiograms, insufficient preoperative echocardiographic data were available in this group of patients for a systematic comparison.

We have a limited number of longitudinal studies in which several echocardiograms were obtained in post-Mustard patients. It appears that certain echocardiographic findings remain constant for a given patient. This includes the position of the baffle within the atrium separating the upper systemic venous limb anteriorly from the pulmonary venous limb posteriorly. The dimensions of structures such as ventricular walls, ventricular cavities, left ventricular outflow tract and pulmonary valve annulus show changes that are appropriate for the patient’s growth.

Patients with aortopulmonary transposition who have undergone Mustard’s procedure cannot be considered to have a normal heart and circulation. It is uncertain whether left ventricular outflow tract obstruction may develop; also, it is not known how long the right ventricle can continue to function as a systemic ventricle, or whether pulmonary hypertension or long term baffle problems will develop. In view of these problems, these patients will require continuous monitoring throughout life. A reliable noninvasive test which can be repeated serially may allow the assessment of these factors and facilitate appropriate timing of repeat cardiac catheterization.

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