Relation of Biventricular Function Quantified by Stress Echocardiography to Cardiopulmonary Exercise Capacity in Adults With Mustard (Atrial Switch) Procedure for Transposition of the Great Arteries

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Background—Mustard repair for transposition of the great arteries (TGA) is frequently associated with impaired systemic (right) ventricular function and sometimes exercise intolerance. We hypothesized that a simple quantitative measurement of ventricular function, during rest and pharmacological stress, could identify abnormalities and predict objective exercise capacity.

Methods and Results—We quantified the performance of systemic and pulmonary (left) ventricles by using echocardiography, at rest and during dobutamine stress, in 27 adults who had undergone Mustard repair for TGA. Systolic and diastolic function of the systemic ventricle were markedly disturbed with respect to pulmonary ventricular function. We also measured exercise capacity by cardiopulmonary exercise testing for peak oxygen uptake. Exercise capacity was significantly predicted by systemic ventricular long-axis excursion both at rest (r=0.66, P<0.001) and at peak dobutamine stress (r=0.53, P=0.006) but not by pulmonary ventricular long-axis excursion at rest (r=0.04) or on stress (r=0.11). Exercise capacity was also predicted by the septal long-axis excursion at rest (r=0.61, P=0.001) but not pulmonary ventricular free wall excursion (P>0.05) or fractional shortening (P>0.05). Peak aortic velocity at maximum dobutamine stress correlated with exercise capacity (r=0.46, P=0.029) but not at rest (r=0.36). Multivariate analysis revealed systemic ventricular long-axis excursion to be the sole significant independent predictor of exercise capacity.

Conclusions—Systemic ventricular function is depressed in most patients with Mustard repair. Quantitative echocardiographic evaluation shows systemic ventricular function to be a key determinant of exercise capacity. (Circulation. 2004;110:1380-1386.)

Key Words: transposition of great vessels ■ echocardiography ■ exercise

After Mustard repair for transposition of the great arteries, systemic (right) ventricular function may be impaired. However, assessing function of this ventricle has proved difficult. Echocardiographic assessment of systemic ventricular function “by eye” is subjective, and formulaic calculation of ejection fraction can be time-consuming and unreliable because of its complex geometry. Other techniques such as MRI have been applied, but they are not widely available.

Since the relation between ejection fraction and exercise capacity is controversial, it might be argued that systemic ventricular function is not an important determinant of exercise capacity in these patients. In the case of the systemic ventricle of a patient with Mustard repair, its unique shape and physiology further adds to the need to broaden our horizon of quantitative measures of ventricular function.

The lack of a gold standard test for the assessment of ventricular function, especially in this group of patients, necessitates the use of an external reference to validate proposed measurements. Cardiopulmonary exercise testing can provide a widely accepted, valid, reproducible, and objective measure of exercise capacity, which is a good candidate for such an external reference point.

The aim of this study was to describe the physiology of the systemic and pulmonary ventricles at rest and during dobutamine stress in patients who had undergone Mustard repair. We sought to quantify not only the conventional ventricular short axis but also the long axis. Our second aim was to validate these echocardiographic measurements by quantifying their relation to objective measurements of exercise capacity by using cardiopulmonary exercise testing.

Methods

From March 2000 to January 2002, 27 patients (29±7 years of age, 12 men) with transposition of the great arteries who had undergone the Mustard procedure were studied. The age at the time of Mustard repair for transposition of the great arteries (TGA) is frequently associated with impaired systemic (right) ventricular function and sometimes exercise intolerance. We hypothesized that a simple quantitative measurement of ventricular function, during rest and pharmacological stress, could identify abnormalities and predict objective exercise capacity.

We quantified the performance of systemic and pulmonary (left) ventricles by using echocardiography, at rest and during dobutamine stress, in 27 adults who had undergone Mustard repair for TGA. Systolic and diastolic function of the systemic ventricle were markedly disturbed with respect to pulmonary ventricular function. We also measured exercise capacity by cardiopulmonary exercise testing for peak oxygen uptake. Exercise capacity was significantly predicted by systemic ventricular long-axis excursion both at rest (r=0.66, P<0.001) and at peak dobutamine stress (r=0.53, P=0.006) but not by pulmonary ventricular long-axis excursion at rest (r=0.04) or on stress (r=0.11). Exercise capacity was also predicted by the septal long-axis excursion at rest (r=0.61, P=0.001) but not pulmonary ventricular free wall excursion (P>0.05) or fractional shortening (P>0.05). Peak aortic velocity at maximum dobutamine stress correlated with exercise capacity (r=0.46, P=0.029) but not at rest (r=0.36). Multivariate analysis revealed systemic ventricular long-axis excursion to be the sole significant independent predictor of exercise capacity.

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repair was 4.1 ± 4.4 years. The time interval between the repair and the study was 25 ± 6 years. No patient had a history of or electrocardiographic evidence for coronary artery disease or systemic hypertension. All patients gave prior written informed consent for this study, which was approved by the local ethics committee.

As a healthy control group, we used data from 22 subjects (38 ± 8 years of age) who had been referred for stress echocardiography and angiography because of atypical symptoms and in whom both tests proved to be normal. All had structurally normal hearts. None had a history of hypertension, diabetes, or typical angina.

Resting Doppler Echocardiography

Transthoracic echocardiography was performed with the use of a Phillips Sonos 5500 echocardiograph interfaced with a multifrequency transducer. With the subject in the left semilateral position, cross-sectional, 2-dimensionally guided, M-mode recordings of the pulmonary (left) ventricular minor axis dimensions were taken at end-diastole (at the onset of q wave of the ECG) and at end-systole (at the onset of the second heart sound on the phonocardiogram), using leading edge methodology. Fractional shortening was calculated as the difference between the two dimensions with respect to the end-diastolic dimension. Systemic ventricular end-diastolic dimension was measured from a frozen 2D image acquired from the same view at the time of the onset of the q wave. Total ventricular long-axis excursion was measured as the excursion of left and right atrioventricular ring motion occurring between the onset of the QRS and the peak innermost point (toward the ventricular cavity) at or after the second heart sound. Post-ejection shortening was measured as the extent of inward movement that occurred after the second heart sound.9

We assessed the reproducibility of the timing measurements of the onset of shortening with respect to the onset of the QRS and the onset of lengthening with respect to S2. Reproducibility expressed as root-mean-square for the intraobserver variability was 11 ms and 14 ms.

Dobutamine Stress Protocol

Dobutamine was administered through an infusion pump (IVAC 770 syringe driver, Alaris Medical System), starting at a rate of 5 μg/kg per minute, increased every 3 minutes by a similar increment to a maximum dose of 40 μg/kg per minute. Stress end points were taken either with the onset of symptoms, arrhythmia (including frequent ventricular extrasystoles), or if the heart rate had reached 85% the predicted target rate for age (220 minus age in years). Blood pressure was recorded at each stage with the use of a Critikon Dinamap monitor (Critikon Inc). All echocardiographic and Doppler parameters were repeated at peak stress, with the use of the same resting setup.

Cardiopulmonary Exercise Testing

Exercise testing was conducted on a treadmill in an air-conditioned room. A Bruce protocol was used, with the addition of a "stage 0," consisting of 3 minutes at a speed of 1 mile per hour with a 5% gradient. Gas exchange was measured breath-by-breath with a heated calibrated pneumotachograph and respiratory mass spectrometer (Amis 2000).

Blood pressure was measured from the right arm, while the patient was standing, before, during, and immediately after the exercise test, with the use of a mercury sphygmomanometer. Heart rate and ECG morphology were continuously monitored and recorded.

The exercise test was performed to the limit of the patient’s physical endurance, with particular exhortation to continue exercise until there was evidence of anaerobic metabolism on gas exchange measurements. Chest pain, sustained arrhythmia, or a fall in systolic blood pressure by >20 mm Hg were also criteria for cessation, but these end points did not occur in any of the studied patients.

Measurements

Doppler Echocardiographic Measurements

Pulmonary ventricular minor axis dimensions were taken at end-diastole (at the onset of q wave of the ECG) and at end-systole (at the onset of the second heart sound on the phonocardiogram), using leading edge methodology. Fractional shortening was calculated as the difference between the two dimensions with respect to the end-diastolic dimension. Systemic ventricular end-diastolic dimension was measured from a frozen 2D image acquired from the same view at the time of the onset of the q wave. Total ventricular long-axis excursion was measured as the excursion of left and right atrioventricular ring motion occurring between the onset of the QRS and the peak innermost point (toward the ventricular cavity) at or after the second heart sound. Post-ejection shortening was measured as the extent of inward movement that occurred after the second heart sound.9

We measured the delay between the onset of electrical activity (QRS) and the onset of shortening in the pulmonary and systemic ventricular free walls (Figure 2). This "q-QS" was previously found to be a sensitive marker of ischemia in patients with coronary artery disease.5,10 We also measured the time interval between the second heart sound and the onset of long-axis lengthening (S2-OL, Figure 2), a measurement similar to isovolumic relaxation time in value despite the difference in their definitions.10

We assessed the reproducibility of the timing measurements of the onset of shortening with respect to the onset of the QRS and the onset of lengthening with respect to S2. Reproducibility expressed as root-mean-square for the intraobserver variability was 11 ms and 14 ms.

Figure 1. Apical 4-chamber view, showing M-mode cursor placed at left, septal, and right AV ring.
Sixteen were in Ability Index I, 8 in II, and 3 in III.11 Sixteen follow-up time. Three had a pacemaker at the time of study. 8 to 24 years (mean, 15) after their original repair, and 3 had balloon/HH11006

Peak systolic, early, and late diastolic velocities were measured from tissue Doppler recordings of the pulmonary and systemic ventricular free wall longitudinal motion taken at the level of the atrioventricular ring. Pulmonary and systemic ventricular filling times were also measured as the time interval from the onset of forward flow to its end. Pulmonary and systemic ventricular ejection times were measured between the onset of forward flow pulse to the onset of the valve closure artifact, respectively.

Cardiopulmonary Exercise Testing
Minute ventilation, oxygen consumption, and carbon dioxide production were calculated online every 10 seconds with the use of a standard inert gas dilution technique (Amis 2000).

Statistical Analysis
For all measured variables, values are expressed as mean± SD. Rest and stress values in patients were compared with those in control subjects through the use of 1-way ANOVA. The systemic and pulmonary ventricles were compared between patients and control subjects by using the same method. A probability value of <0.05 was considered significant. The Pearson product-moment method was used for calculating correlation coefficients, and multivariate regression was performed by the stepwise method with F-to-enter of 4.

Results
Six patients had undergone redo Mustard operation 8 to 24 years (mean, 15) after their original repair, and 3 had balloon dilation for the venous pathway obstruction during recent follow-up time. Three had a pacemaker at the time of study. Sixteen were in Ability Index I, 8 in II, and 3 in III.11 Sixteen were in New York Heart Association class I, 10 in II, and 1 in III. Twenty-four patients had a trace or mild degree of tricuspid regurgitation, and the remaining 3 had moderate tricuspid regurgitation.

At peak dobutamine stress, no patient had chest pain, 1 had shortness of breath, and another had lightheadedness. The remaining 25 completed the protocol without any significant limiting symptoms. Twenty-five patients performed cardiopulmonary exercise testing; 1 could not tolerate the mouthpiece, and 1 declined to attempt the test.

Three patients had pacemakers: 1 AAIR and 2 DDR. Only one was receiving ventricular pacing stimuli during the study; timing measurements were not made in him.

Systemic Ventricular Function
During systole, the patients with Mustard repair had a longer delay in the onset of shortening of the ventricular free wall, a smaller long-axis excursion, and lower systolic velocity and ventricular outflow velocity. Moreover, their stress-induced increment in long-axis excursion, systolic velocity, and ventricular outflow velocity was significantly smaller than in the control subjects (Table 1, interaction column).

Diastole also was disordered in patients, with delayed onset of long-axis lengthening, reduced myocardial velocities, and higher early trans–AV valve flow velocity. The effect of stress in diastole was not significantly different between patients and control subjects. At rest, 7 patients had no detectable "A" filling wave. During stress, this number rose to 15.

Pulmonary Ventricular Function
Pulmonary ventricular function in patients with Mustard repair was also abnormal (Table 1). The time from q wave to the onset of ventricular shortening failed to shorten on stress. Long-axis excursion was lower, with higher peak outflow tract velocity, reflecting an element of outflow tract obstruction. Outflow tract obstruction, defined as systolic anterior movement of the mitral valve causing outflow tract velocities to exceed 3.5 m/s, occurred in 10 of 27 patients (1 at rest and 9 at peak stress).

During diastole, lengthening began prematurely, tissue Doppler early diastolic velocity and early diastolic filling velocity were significantly elevated, and tissue Doppler late diastolic velocity was abnormally low. The patients showed a smaller stress-induced increment in late diastolic velocity than did the control subjects.

Incoordination (Post-Ejection Shortening)
At Rest
Seven of 24 patients presented with incoordination (post-ejection shortening) of the long axis (4 had it at the septum at the same time) in the absence of left bundle-branch block.

During Stress
Incoordination worsened in 5 of the 7 patients and newly developed in 12 other patients (3 septal site, 5 systemic ventricle, 4 at both septal and systemic ventricular free wall) (Figure 3). Thirteen of the total 19 patients who had incoordination with stress showed abnormal ST-segment shift on the 12-lead ECG. None of the control subjects had incoordination at rest, nor did any have it during stress.

Exercise Capacity in Patients With Mustard Repair
Exercise capacity was significantly depressed in many patients compared with normal reference values, but the range was wide (Figure 4). Exercise data are shown in Table 2.

Predictors of Exercise Capacity in Patients With Mustard Repair
Long-Axis Excursion
Exercise capacity correlated significantly with systemic ventricular free wall excursion, both at rest (r=0.66, P<0.001, Figure 5) and during dobutamine stress (r=0.53, P=0.006). It also correlated with septal long-axis excursion at rest.
TABLE 1. Comparison of Ventricular Function Between Patients and Controls

<table>
<thead>
<tr>
<th></th>
<th>Controls</th>
<th>Patients</th>
<th>Patient Versus Control</th>
<th>Stress Versus Rest</th>
<th>Interaction Between Patient/Control and Stress/Rest</th>
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<td>Mean ± SD</td>
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<tr>
<td>Systemic ventricle</td>
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<td>Systolic physiology</td>
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<tr>
<td>Q to onset of shortening, ms</td>
<td>89.1±17.2</td>
<td>55±12.2</td>
<td>119.6±27.1</td>
<td>95.6±45.4</td>
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<td>Long-axis excursion, cm</td>
<td>1.55±0.22</td>
<td>1.85±0.2</td>
<td>1.03±0.33</td>
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<td>Ventricular systolic velocity, cm/s</td>
<td>8.5±1.8</td>
<td>14.6±3.9</td>
<td>8.4±3.1</td>
<td>10.4±3.7</td>
<td>7.2</td>
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<tr>
<td>Peak ventricular outflow velocity, m/s</td>
<td>1.1±0.13</td>
<td>2.1±0.21</td>
<td>0.99±0.15</td>
<td>1.49±0.32</td>
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<td>Time from S2 to onset of lengthening, ms</td>
<td>65.5±13.7</td>
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<td>90.4±39.7</td>
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<td>Tissue Doppler early diastolic velocity, cm/s</td>
<td>13.4±3.6</td>
<td>15.1±2.9</td>
<td>10.8±3.5</td>
<td>12.8±3.5</td>
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<td>Tissue Doppler late diastolic velocity, cm/s</td>
<td>10.9±2.6</td>
<td>14±3.3</td>
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<td>Doppler E wave velocity, cm/s</td>
<td>62.7±15.5</td>
<td>82.3±17.2</td>
<td>79.6±29.6</td>
<td>95.4±28.7</td>
<td>6.5</td>
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<tr>
<td>Pulmonary ventricle</td>
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<td>Systolic physiology</td>
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<td>Q to onset of shortening, ms</td>
<td>82.3±21.9</td>
<td>48.6±11.7</td>
<td>68.1±24</td>
<td>61.9±14.4</td>
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<tr>
<td>Long-axis excursion, cm</td>
<td>2.37±0.16</td>
<td>2.8±0.3</td>
<td>1.73±0.44</td>
<td>2.1±0.47</td>
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<td>Ventricular systolic velocity, cm/s</td>
<td>11.2±1.8</td>
<td>20.1±2.6</td>
<td>12.5±5.2</td>
<td>21.6±4.8</td>
<td>2.0</td>
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<tr>
<td>Peak ventricular outflow velocity, m/s</td>
<td>0.76±0.11</td>
<td>1.6±0.47</td>
<td>1.46±0.92</td>
<td>2.72±1.29</td>
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<td>Diastolic physiology</td>
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<tr>
<td>Time from S2 to onset of lengthening, ms</td>
<td>28.6±22.7</td>
<td>20±18.5</td>
<td>10±34.6</td>
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<tr>
<td>Tissue Doppler early diastolic velocity, cm/s</td>
<td>11.6±2.7</td>
<td>14.1±3.5</td>
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<tr>
<td>Tissue Doppler late diastolic velocity, cm/s</td>
<td>11.5±2.5</td>
<td>19.9±4.9</td>
<td>10±4.4</td>
<td>13.4±3.3</td>
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<td>Doppler E wave velocity, cm/s</td>
<td>43.8±12</td>
<td>60±14.1</td>
<td>81.9±23.8</td>
<td>97.7±26.3</td>
<td>59.9</td>
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</table>

(r=0.61, P=0.001) but not with pulmonary ventricular excursion (r=0.042) or with fractional shortening (r=0.13).

Doppler Flow Velocity
Peak aortic velocity on stress correlated with exercise capacity (r=0.46, P=0.03) although resting values did not (r=0.36).

Multivariate Analysis
A stepwise multivariate regression analysis was performed to identify which echocardiographic measurements showed independent association with exercise capacity. Systemic ventricular free wall long-axis excursion was the strongest correlate of exercise capacity and therefore entered the model first. In its presence, no other variable added significant predictive value; therefore it alone remained in the multivariate analysis (P<0.005).

Discussion
In this quantitative study of ventricular function during dobutamine stress echocardiography applied to the long axis of the heart, we have found that systemic ventricular function is substantially impaired in patients with the Mustard procedure. Second, objective exercise capacity was impaired in many of such patients, even though they considered themselves asymptomatic or only minimally symptomatic. Third, systemic ventricular function, as quantified by echocardiographic long-axis measurement, is the key correlate of objective exercise capacity.

Ventricular Function
In patients with structurally normal hearts, the conventional approach to assessment of systemic ventricular function is to measure ejection fraction. However, attempting to extend this to the Mustard scenario has proved problematic. The geometry of the right ventricle makes it difficult to measure ejection fraction by echocardiography, which has prompted simple visual estimation or more complex and expensive techniques such as radionuclide or magnetic resonance ventriculography.12,13

In contrast, simple measurement of long-axis excursion gives a rapid and unambiguous measurement,14-16 which has proven utility as a measure of left ventricular function in patients with coronary artery disease, valve disease, and heart failure.17,18 The technique is equally straightforward for the right ventricle and is especially valid because the main bulk of right ventricular myocardial fibers are arranged longitudinally.

Tissue Doppler also identified depressed early and late diastolic myocardial velocities in patients with Mustard repair. Despite this, early diastolic filling velocities were...
abnormally high. Although there was an age difference between groups, age alone could not explain this combination of findings; rather, there may be diastolic restriction.19

Using this quantitative approach, we found that 80% of patients had significantly depressed systemic ventricular long-axis function with stress. This broadly agrees with findings from radionuclide and MRI studies.12,20 However, since there is no gold standard among imaging techniques for assessing the abstract concept of “function,” validity of the measurements must be established by reference to an external, nonimaging, modality.

Exercise Capacity
Cardiopulmonary exercise testing with measurement of peak oxygen uptake is an objective, valid, and reproducible method of grading exercise capacity.21,22 Even though the majority of patients in the group were asymptomatic or almost so, many had marked exercise impairment. This paradox may result from the lifelong nature of the condition, which allows the patient to downgrade their expectations of “normal” activity. Moreover, normal everyday activity (other than sports) rarely approaches peak exercise levels, and thus depressed peak exercise capacity may not encroach much on daily life. Nevertheless, in patients (without congenital heart disease) with the potentially analogous condition of systemic left ventricular dysfunction, peak exercise capacity is an important predictor of survival.1

Ventricular Function and Exercise Capacity
Previous data on the relation of ventricular function and exercise capacity in patients with Mustard repair have been conflicting.3,23,24 Several measures of the systemic ventricular physiology were linked to exercise capacity. Multivariate analysis demonstrated that the key information that they provided—reduced systemic ventricular long-axis function—was com-
mon between them, so that when one variable was in the model, the others did not contribute significantly.

This suggests that central hemodynamics play a powerful role in limiting exercise capacity in patients who have had the Mustard operation. This relation may be stronger than the corresponding relation in patients with chronic heart failure caused by left ventricular disease because of more frequent coexisting extracardiac pathologies in the latter, who are typically much older.

Measurement of systemic ventricular free wall motion is simple and unambiguous and is found to be the measurement most closely related to exercise capacity in these patients, whether it is measured at rest or on stress. This supports the concept that the overwhelmingly important limiting factor to exercise capacity in these patients is disruption of purposeful systolic function of the ventricle supporting the systemic circulation.

Evidence of Ischemic Pattern of Ventricular Function

Shortening of the long axis normally ends before the end of ejection. When there is myocardial ischemia, shortening may continue after ejection (incoordination or “post–ejection shortening”). This phenomenon is sufficiently predictive of coronary artery disease (in patients without congenital heart disease) in that it can be used as an objective end point suggesting inducible ischemic dysfunction during diagnostic stress echocardiography.10,9,25–27

Thirteen of our patients showed post–ejection shortening of the long axis, in association with abnormal ST-T segments changes on the ECG at peak stress. Although ECG abnormalities may be difficult to interpret, the post–ejection shortening is characteristic of ischemic dysfunction.9 These findings are in accord with previous studies that used radionuclide perfusion imaging, which have identified regional inducible ischemia in the systemic right ventricle in patients with Mustard repair.12

Inducible ischemia does not necessarily require epicardial coronary artery stenosis because incoordinate ventricular behavior in early diastole may compromise regional filling of coronary arteries and their capillaries, resulting in local ischemia. High systemic ventricular afterload also increases wall stress and therefore may further compromise subendocardial myocardial perfusion. Thus, the very hypertrophy of the right ventricle that permits it to sustain a high pressure circulation could be simultaneously increasing the myocardial oxygen demand and interfering with normal oxygen supply demand balance. Finally, systemic ventricular chamber dilation, commonly seen in Mustard repair, will also increase chamber wall tension and potentially compromise further myocardial perfusion.

Abnormal Pulmonary Ventricular Function

The pulmonary ventricle in patients with Mustard repair has an earlier onset of relaxation and a higher early diastolic myocardial velocity and early filling velocity than the pulmonary ventricle of control subjects. This may represent the morphological left ventricle, originally evolved to suit the high afterload of the systemic circulation, being applied to the much lower resistance of the pulmonary vascular bed. Even though the result is abnormally high pulmonary ventricular velocities, it does not prevent exercise capacity being impaired.

Study Limitations

We set out to address systemic right ventricular function by using easily measurable quantitative parameters requiring widely available equipment. We are therefore unable to compare the relative merits of different imaging modalities (such as radionuclide or magnetic resonance) against those of echocardiography. However, we found that in every patient, we were able to measure the echocardiographic parameters without ambiguity or contraindication and that systemic ventricular long-axis physiology related usefully to exercise capacity. The healthy subjects were older than the patients. However, the age difference was not extreme and should not cause lower values for cardiac function. Thus, these abnormalities found in patients with Mustard repair are likely to represent a significant difference from normal.

Conclusions

Systemic right ventricular function in patients with Mustard repair is impaired at rest and during pharmacological stress (even among asymptomatic ones) and appears to limit exercise capacity. Pulmonary ventricular function is also impaired but to a lesser degree. Excursion of the systemic ventricular free wall is the dominant predictor of exercise capacity and should be considered for the routine periodic assessment of these patients.

References


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