Exercise Performance After Repair of Anomalous Origin of the Left Coronary Artery From the Pulmonary Artery

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Eleven patients underwent exercise testing after operative repair of anomalous origin of the left coronary artery from the pulmonary artery. Five patients repaired after 2 years of age comprised a childhood surgery group, and six patients repaired before 2 years of age comprised an infant surgery group. All patients were exercised using either a treadmill or electronically braked bicycle with simultaneous thallium 201 scintigraphy. Oxygen consumption, carbon dioxide production, pulmonary functions, and electrocardiogram were all monitored continuously. Pulmonary reserve was normal in all patients. Based on heart rate reserve, respiratory exchange ratio, and oxygen-consumption response to work load, two patients in the infant surgery group stopped exercise before achieving maximum aerobic capacity. All remaining patients achieved their maximum aerobic capacity. There was no difference in work rate or oxygen consumption during exercise between the infant and childhood surgical group. Four patients (two in each surgical group) had an impaired chronotropic response to exercise. Three of these four patients demonstrated perfusion defects by thallium scintigraphy. Thallium scintigraphy was normal in all remaining patients. Electrocardiographic abnormalities were noted in seven of 11 patients having ventricular arrhythmias or ST segment depression. It is concluded from this study that exercise performance after repair of anomalous origin of the left coronary artery from the pulmonary artery is not affected by the age at which surgery is performed. Exercise is frequently associated with electrocardiographic evidence of abnormal myocardial perfusion despite frequently negative simultaneous 201TI scintigraphy. (Circulation 1990;81:1287–1292)

Anomalous left coronary artery arising from the pulmonary artery is a rare defect explaining fewer than 0.1% of new cases of congenital heart disease. It frequently results in congestive heart failure secondary to myocardial ischemia and subsequent infarction in early infancy.1 The need for surgery, the timing of surgery, and the type of operation for these patients have been debated for years. There seems to be a general consensus that establishing a two coronary artery system is preferable to assure adequate myocardial perfusion.1–7

To aid in the evaluation of cardiac function of patients with anomalous origin of the left coronary artery from the pulmonary artery, this study evaluated the exercise performance of a group of patients who had undergone surgical repair of this defect. We evaluated both myocardial performance and, using thallium 201 scintigraphy, postoperative myocardial perfusion.

Methods

Population

Eleven children and adolescents, two boys and nine girls, comprised the study group. The population was divided into two groups. Patients 1–5 underwent the operation at more than 2 years of age. Patients 6–11 were operated on when less than 2 years old (Table 1).

All patients had undergone operative repair before the study. Two patients also underwent mitral valve replacement for severe mitral regurgitation. Surgical procedures consisted of a saphenous vein graft from the ascending aorta to the left main coronary artery in four patients. Three patients had a left subclavian artery graft from the ascending aorta to the left main coronary artery. Two patients had an aortopulmonary window created with an intrapulmonary baffling of the left coronary ostia to the proximal ascending aorta. One patient underwent ligation of the left coronary artery as the only procedure.

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Both patients with mitral valves were receiving coumadin. No patient was receiving any other medication. All patients were New York Heart Association functional class I at the time of exercise testing.

**Procedures**

**Catheterization.** Preoperative left ventricular end-diastolic volume indexed to body surface area and left ventricular ejection fraction were calculated for each patient. The same measurements were repeated on those patients who underwent postoperative cardiac catheterization.

**Echocardiogram.** All patients underwent a two-dimensional and M-mode echocardiogram within 1 month before exercise testing. Left ventricular shortening fraction was calculated and compared with preoperative shortening fractions on those patients where preoperative echocardiograms were available.

**Pulmonary testing.** Immediately before exercise testing, the patients underwent pulmonary function testing, consisting of maximum inspiratory and expiratory flow-volume loops and maximum voluntary ventilation (MVV).

**Exercise protocol.** All patients were exercised to maximum voluntary effort. Eight were tested with an electronically braked bicycle ergometer. The protocol consisted of 3 minutes of unloaded cycling followed by 1-minute incremental increases in the work rate, calculated to achieve the patient's predicted maximum work rate in 10–12 minutes. Three patients were too small for cycle testing and underwent treadmill testing using either the Bruce protocol (two patients) or a 1-minute incremental protocol similar to the bicycle protocol (one patient). One minute before exercise termination, 0.03 mCi of $^{201}$Tl was injected intravenously into an antecubital vein.

**Electrocardiographic monitoring.** A 12-lead electrocardiogram (ECG) was obtained in the sitting and standing position before exercise testing. Leads II, aVF, and V5 were monitored continuously. A 12-lead ECG was obtained at each 1 minute of exercise and at 3 and 5 minutes into the recovery period. A signal average beat for leads II, aVF, and V5 was obtained each minute. Significant ST segment depression was defined as a more-than-1.0-mm level or downward sloping depression occurring more than 60 msec after the J-point in the signal-averaged beats.

**Metabolic measurements.** Minute oxygen consumption ($\text{VO}_2$) and carbon dioxide production were monitored with a breath-by-breath method using a commercially available metabolic cart. Respiratory exchange ratio, minute ventilation ($\text{VE}$), and tidal volume were monitored breath by breath. The ventilator equivalents of oxygen and carbon dioxide ($\text{VE}/\text{VO}_2$, $\text{VE}/\text{VCO}_2$) were monitored continuously and used to determine onset of ventilator anaerobic threshold (VAT) by established techniques.

**Criteria for evaluating reason for exercise termination.** Pulmonary reserve was believed to be nonlimiting if the ratio of maximum achieved $\text{VE}$ to the preexercise MVV was 0.85 or less. Attainment of maximum aerobic capacity was defined as achieving a respira-
TABLE 2. Exercise and Myocardial Perfusion Data

<table>
<thead>
<tr>
<th>Patient</th>
<th>Work rate (W)</th>
<th>Work rate (% pred #)</th>
<th>Max HR (beats/min)</th>
<th>Max VO₂ at anaerobic threshold (ml/kg/min)</th>
<th>%Max VO₂ at anaerobic threshold</th>
<th>Max RER</th>
<th>Max VO₂/Vt/MVV</th>
<th>Electrocardiographic changes</th>
<th>Thallium scintigraphy</th>
<th>Graft patency at catheter</th>
</tr>
</thead>
<tbody>
<tr>
<td>Childhood surgery</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>275</td>
<td>102</td>
<td>182</td>
<td>50</td>
<td>21</td>
<td>42</td>
<td>1.24</td>
<td>0.82</td>
<td>Frequent PVCs with exercise</td>
<td>Negative</td>
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<tr>
<td>2</td>
<td>240</td>
<td>113</td>
<td>167</td>
<td>38</td>
<td>18</td>
<td>47</td>
<td>1.38</td>
<td>0.75</td>
<td>Frequent PVCs, nonsustained V-tach</td>
<td>Negative</td>
</tr>
<tr>
<td>3</td>
<td>130</td>
<td>72</td>
<td>188</td>
<td>22</td>
<td>9</td>
<td>41</td>
<td>1.21</td>
<td>0.63</td>
<td>None</td>
<td>Negative</td>
</tr>
<tr>
<td>4</td>
<td>120</td>
<td>74</td>
<td>192</td>
<td>26</td>
<td>...</td>
<td>...</td>
<td>1.32</td>
<td>0.77</td>
<td>Atrial flutter, nonsustained V-tach</td>
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</tr>
<tr>
<td>5</td>
<td>92</td>
<td>88</td>
<td>175</td>
<td>23</td>
<td>15</td>
<td>77</td>
<td>1.14</td>
<td>0.54</td>
<td>2-mm ST segment depression in V5 and aVF</td>
<td>Globally dilated LV</td>
</tr>
<tr>
<td>Mean</td>
<td>171</td>
<td>89</td>
<td>180</td>
<td>31</td>
<td>15</td>
<td>51</td>
<td>1.25</td>
<td>0.7</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Infant surgery</td>
<td></td>
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<td></td>
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<td>109</td>
<td>184</td>
<td>31</td>
<td>...</td>
<td>...</td>
<td>1.35</td>
<td>0.64</td>
<td>3-mm ST segment depression in V5 and aVF</td>
<td>Negative</td>
</tr>
<tr>
<td>7</td>
<td>80</td>
<td>98</td>
<td>178</td>
<td>39</td>
<td>15.6</td>
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<td>1.21</td>
<td>0.59</td>
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<td>Negative</td>
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<tr>
<td>8</td>
<td>120</td>
<td>74</td>
<td>198</td>
<td>26</td>
<td>13</td>
<td>50</td>
<td>1.26</td>
<td>0.87</td>
<td>None</td>
<td>Negative</td>
</tr>
<tr>
<td>9</td>
<td>75</td>
<td>77</td>
<td>152</td>
<td>20</td>
<td>...</td>
<td>...</td>
<td>1.02</td>
<td>0.56</td>
<td>2-mm ST segment depression in V5 and aVF</td>
<td>Negative</td>
</tr>
<tr>
<td>10</td>
<td>51</td>
<td>67</td>
<td>163</td>
<td>28</td>
<td>...</td>
<td>...</td>
<td>1.13</td>
<td>0.66</td>
<td>2-mm ST segment depression in V5 and aVF</td>
<td>Negative</td>
</tr>
<tr>
<td>11</td>
<td>100</td>
<td>69</td>
<td>176</td>
<td>27</td>
<td>14.7</td>
<td>54</td>
<td>1.21</td>
<td>0.51</td>
<td>None</td>
<td>Negative</td>
</tr>
<tr>
<td>Mean</td>
<td>87†</td>
<td>82</td>
<td>175</td>
<td>29</td>
<td>14</td>
<td>48</td>
<td>1.20</td>
<td>0.64</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

% pred, percent of predicted normal; HR, heart rate; VO₂, minute oxygen consumption; RER, respiratory exchange ratio; Max Vt, minute ventilation at maximum exercise; MVV, maximum voluntary ventilation; PVC, premature ventricular contraction; V-tach, ventricular tachycardia.


†p<0.05, different from childhood surgery group.
tory exchange ratio greater than 1.12 with the graphs of VE/VO$_2$ and VE/VCO$_2$, indicating a respiratory pattern consistent with significant metabolic acidosis. Maximum heart rate and plateauing of oxygen consumption in response to increasing work rate were used as additional confirmation of achieving maximum aerobic capacity.

**Thallium 201 scintigraphy.** Immediately after exercise testing, the patients were scanned in 35–45° left anterior oblique, anterior, and lateral views. Redistribution scans using the same views were obtained 4 hours after exercise. Scan results were interpreted by one of the authors without knowledge of exercise performance.

**Statistical analysis.** Differences between the infant and childhood surgery groups and differences within each group preoperatively and postoperatively were analyzed using the Student’s two-tailed $t$ test. A $p$ value less than 0.05 was considered significant.

**Results**

Table 1 displays the ages of the patients at the time of diagnosis, surgery, postoperative cardiac catheterization, and exercise study. The childhood surgery group was significantly older than the infant surgery group at both age of diagnosis and surgery as well as the time of exercise study.

**Catheterization and Echocardiographic Data**

Preoperative and postoperative angiographically determined left ventricular end-diastolic volume indexed for body surface area and ejection fraction are displayed in Table 1. Only two patients in the childhood surgery group have undergone postoperative cardiac catheterization. There was no difference between the two groups either preoperatively or postoperatively. There was, however, a significant decrease in ventricular volume and an increase in ejection fraction for the entire patient population postoperatively, as compared with their preoperative values.

Similarly, echocardiography showed a significant increase in left ventricular shortening fraction for the entire population in the postoperative state (Table 1). Although the shortening fraction in the infant group postoperatively was somewhat greater than in the childhood surgery group (as was the angiographically derived ejection fraction), this was not significant. No patient had any wall motion abnormality on the resting echocardiogram.

**Exercise Performance**

Table 2 summarizes the exercise performance of the study population. No patient in either group seemed to have a respiratory limitation of exercise based on their VE-to-MVV ratio (Table 2). Patient 8 had a slightly elevated VE-to-MVV ratio but this was believed to be because of a poor effort in obtaining the resting MVV. Her vital capacity and her inspiratory and expiratory flow-volume loops were normal. Two patients in the infant surgery group (patients 10 and 11) had their exercise testing terminated because of the onset of acute hyperventilation. This was marked by a rapid increase in the respiratory exchange ratio with a decrease in the end-tidal PCO$_2$. In both cases, this was preceded by an erratic ventilation pattern. The remaining four patients in the infant group and all patients in the childhood group terminated exercise after achieving maximum aerobic capacity.

Comparison between the childhood and infant surgery groups showed only work rate to be significantly different. This was because of the older age of the childhood group at the time of testing, and they would, therefore, be expected to have a higher absolute work rate. When work rate was expressed as a percentage of the predicted maximum for age, height, weight, and sex, there was no difference between the two groups (Table 2).

Maximum heart rate was significantly below the predicted values for the two patients (patients 9 and 10) in the infant surgery group who had submaximal testing. Additionally, significant chronotropic impairment was found in two patients in the childhood group (patients 2 and 5) and two in the infant group (patients 7 and 11) who clearly reached maximum aerobic capacity based on their respiratory and metabolic data.

Maximum oxygen consumption was similar for the two operative groups. Maximum oxygen consumption was reduced in two patients (patients 4 and 5) of the childhood group (Table 2). Note that patient 3 was markedly obese (99 kg) and that her maximum VO$_2$ was 97% of predicted for age and sex when corrected to her ideal body weight. In the infant group, maximum oxygen consumption was reduced in two of the patients (patients 8 and 11) who achieved maximum aerobic capacity.

Oxygen consumption at the onset of VAT was decreased in all patients in whom it could be accurately measured (Table 2). It was also decreased when expressed as a fraction of the patient’s maximum oxygen consumption except for patients 4 and 11 (Table 2).

**Electrocardiographic Data**

Three of the five patients in the childhood surgery group had arrhythmias consisting of PVCs, ventricular tachycardia, and atrial flutter. One additional patient had ST segment depression in the inferior and lateral leads (Table 2). No patient in the infant surgery group had arrhythmias but three patients had ST segment depression (Table 2).

**Thallium Scintigraphy**

Two of the patients (patients 4 and 5) in the childhood surgery group had abnormal thallium scans. Both of these patients had associated electrocardiographic abnormalities. Neither had undergone a postoperative catheterization (Table 2). One patient (patient 8) in the infant group had an abnormal thallium scan and was noted to have a patent graft on previous postoperative cardiac catheterization (Table 2).
Discussion

Management of anomalous origin of left coronary artery from the pulmonary artery has remained controversial. Although most now agree that establishing a two coronary artery system is preferable, the timing and type of operation remain under debate.2–7 Because of the small number of patients in any series, the long-term evaluation of these patients undergoing various operative procedures is very difficult. Previous exercise studies have limited reporting to electrocardiographic changes, thallium perfusion imaging, or both,12–14 and did not include any patients repaired in infancy.

Cardiac Catheterization and Echocardiographic Data

Age at surgical repair did not appear to be a factor in either the cardiac catheterization or echocardiographic measurements of resting left ventricular function. Comparing these findings, as well as the exercise performance and myocardial perfusion, with the presence of a patent graft is difficult because three of the patients had not received postoperative catheterizations, and most of the remaining catheterizations were remote from the time of exercise testing.

There does not seem to be any relation to graft patency and evidence of exercise-induced myocardial ischemia. It should be noted that patient 3, who had ligation of her artery, had a normal maximum oxygen consumption and no evidence of myocardial ischemia. This would suggest that coronary collateral supply continues to be an important determinant of myocardial perfusion and ventricular performance in at least some patients.

Exercise Performance

Working capacity and maximum oxygen consumption were similar for the childhood and infant surgery groups. Four patients had only mild limitations. This would suggest age at surgery is not a factor in later cardiac performance during exercise. Similarly, chronotropic impairment seems to occur equally in both surgical groups. Although chronotropic impairment can reflect myocardial ischemia in adults with coronary artery disease,9 it has not been previously reported in patients with anomalous origin of the left coronary artery from the pulmonary artery. The presence of electrocardiographic changes or positive thallium scans in three of these four patients tends to support the idea that compromised coronary blood flow might have resulted in this chronotropic impairment.

VO₂ at VAT was low in all seven patients in whom it could be clearly measured when compared with the previously published normal data of 30–35 ml/kg/min for children and adolescents.8,15 Although the absolute values of the VO₂ at VAT are low for patients 4 and 11, when these values are expressed as a percentage of their low maximum VO₂, they are within the normal range (Table 2).8,15 This is compatible with exercise performance in the presence of diminished cardiovascular reserve from ischemic heart disease.10

The values for the other four patients are low both as absolute values and when expressed as percentages of their achieved maximum VO₂.8,15 A possible explanation might be the sedentary lifestyle of these patients. VO₂ at VAT is probably a better marker of cardiovascular fitness than maximum VO₂ and is more readily influenced by changes in the level of physical activity.10,15 These patients’ inactive lifestyles can be reflected by their low VO₂ at VAT despite a normal maximum VO₂.

Electrocardiographic and Thallium Scintigraphy Data

Abnormal electrocardiographic response to exercise was seen in seven of the 11 patients; only patients in the childhood surgery group had ventricular arrhythmias. The numbers are too small, however, to draw any conclusions about the possible differences between the two groups.

There was not any correlation within either surgical group between electrocardiographic changes and the presence of abnormal thallium scans. In fact, five patients had arrhythmias or ST segment changes with normal thallium scans.

In previous series, Finley et al12 noted persistent ST segment depression in two of their three postoperative patients despite negative thallium scans, and suggested undetected subendocardial ischemia as a possible mechanism. Moodie et al13 reported a series of 10 patients operated on as adults, all of whom had negative electrocardiographic findings and negative thallium scans. They suggested that thallium scans could accurately identify ischemia if exercise and resting scans were performed and compared with each other.13 El-Said et al14 did not perform thallium scans on the group of patients they exercised. Only one patient had abnormal ST segment depression and had an occluded coronary artery graft.14

Using exercise and reperfusion scanning, our data tend to support Finley’s12 contention that residual exercise-induced ischemic changes not detected by thallium scans persist postoperatively. The reasons for this poor correlation of electrocardiographic and thallium scintigraphy findings are not clear. Sub-maximal stress of the coronary supply is an unlikely reason because all but two of the patients in our study clearly achieved maximum aerobic effort. A more likely explanation might be the limited ability of thallium scintigraphy to detect isolated subendocardial ischemia. This is supported by the findings that those three patients with cardiac dysfunction severe enough to result in diminished maximum VO₂ are also the three patients with abnormal thallium scans. Thallium scintigraphy, therefore, might be too insensitive to detect persistent perfusion defects in those patients with normal aerobic capacity. This is an important limitation because, as the two patients who developed ventricular tachycardia demonstrate, these patients can still have potentially life-threatening responses to exercise despite their nor-
normal maximum $\dot{V}O_2$. We would, therefore, recommend close observation of patients with normal exercise capacity and negative thallium scans if they demonstrate any electrocardiographic abnormalities.

References

**KEY WORDS** • thallium scintigraphy • myocardial perfusion
Exercise performance after repair of anomalous origin of the left coronary artery from the pulmonary artery.

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