Muscular Strength and Function in Patients With Cystic Fibrosis*

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Background: For 20 years, physical activity has been an important component in the treatment of cystic fibrosis (CF) patients in Sweden. Data concerning physical performance in terms of muscular strength in these patients are limited.

Objective: To compare muscular strength and function in patients with CF with those aspects in a healthy control group (CG).

Design: Thirty-three patients with CF (16 women) aged 16 to 35 years and 20 healthy individuals matched for age and gender were included in the study. All participants had undertaken regular physical training two to three times per week. The following tests were performed: vertical jumping ability; hand-grip strength; abdominal strength; arm/shoulder strength; quadriceps muscle strength; and a functional test of leg muscle endurance.

Results: Patients with CF showed decreased muscle strength and function compared to control subjects (women: maximal hand-grip strength in the right [p = 0.02] and the left hand [p = 0.001]; sustained hand-grip strength in the left hand [p = 0.002]; and in leg muscle endurance [p = 0.02]; men: the number of sit-ups performed within 30 s [p = 0.03]; and left leg isokinetic quadriceps strength at 180° per second [p = 0.02]). The differences were not related to pancreatic or pulmonary function. There was no significant difference between the CF group and the CG in any other test results.

Conclusions: Our study showed few differences in muscular performance between patients with CF and healthy control subjects. Both groups had regular moderate-to-high activity levels. Further studies are needed to evaluate whether the small but significant differences might be related to metabolic abnormalities in skeletal muscles in CF patients.

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Key words: gender; hand-grip strength; lung function; muscle tests; physical activity

Abbreviations: BMI = body mass index; CF = cystic fibrosis; CG = control group; MVC = maximal voluntary contraction; PI = pancreas insufficient; PS = pancreas sufficient; SMVC = sustained maximal voluntary contraction

Cystic fibrosis (CF) is a chronic hereditary disease that influences many systems in the body, mainly the respiratory, digestive, and reproductive systems. Since the 1980s, routine CF treatment in Sweden has included physical training and encouragement to participate in different athletic activities. Treatment focuses on physical activity from an early age, such as trampoline jumping from the age of 1.5 years. Antibiotics and nutrition regimens are important components of treatment, as well as individualized chest physiotherapy. Muscular strength and function are essential in daily life as well as during sports activities. Patients with CF have been reported to have less muscular strength and to be weaker than healthy individuals. Most, but not all, of these studies have been performed in stable patients with moderate-to-severe disease. Some studies have included children, and usually only a few muscles have been tested. Very few researchers have assessed their studies results from a gender perspective, and in only one study were athletic CF children compared to healthy control subjects according to gender.

The ideal type, intensity, frequency, and duration...
of physical activity for patients with CF is unknown. Hitherto, instructions have been given that are similar to those given to healthy individuals, as follows: do any kind of physical training three times per week and for at least 30 min in each session. Some patients prefer muscle-resistance training, while others prefer endurance training, and many want to participate in different sports. Endurance training affects the cardiorespiratory systems, increasing heart and respiration rates. Adolescent CF patients often decrease their training over time. Whether this reduction is linked to psychological factors or to true physical impairment in endurance has not yet been studied.

It is well-known that there are differences in muscular strength and function between healthy men and women. The aims of this study were to measure muscular strength and function in our young adult CF population and to make comparisons with a healthy control group (CG) with regard to gender.

**Materials and Methods**

**Subjects**

During 2002 and 2003, 55 patients aged 16 to 35 years regularly attended the West Swedish CF Center in Göteborg. The CF diagnosis was verified by sweat testing (Cl, > 60 mEq/L), clinical criteria, and genetic analysis. Four patients received diagnoses after the age of 15 years. Eight patients were excluded from the study due to lung transplantation (two women), because they were awaiting lung transplantation (two women and one man), they were receiving oxygen supplementation during sleep or exercise (one man), and pregnancy (two patients). Fourteen patients declined to participate in the study due to lung transplantation (two women), because they had other severe mutations. A total of 24 patients had severe mutations and were PI (11 women). Nine patients had mild genotypes and were pancreas sufficient (PS) [five women], Twenty-two patients were chronic *Pseudomonas aeruginosa* colonization. Six patients had CF-related diabetes mellitus, six patients had mild liver involvement verified by liver biopsy, and one patient had liver cirrhosis. These seven patients had been treated with ursodeoxycholic acid for years. One patient received oral corticosteroids periodically. None of the patients received inhaled corticosteroids.

The body composition was evaluated by dual x-ray absorptiometry in 28 patients with CF (15 women), and lean body mass and fat mass were normal: lean body mass for women, 66% of body weight (SD, 6.0% of body weight); lean body mass for men, 79% of body weight (SD, 5.6% of body weight); fat mass for women, 30% of body weight (SD, 6.2% of body weight); and fat mass for men, 16% of body weight (SD, 5.8% of body weight). No differences between PI and PS patients were observed (E. Gronowitz, Lic Med; personal communication; May 2004). The five patients (one woman) who were not included in those assessments for technical reasons did not differ from the others in terms of anthropometry. Twenty healthy participants matched for age and gender (eight women) served as a CG (Table 1). Two women with CF were secondi para, as were the corresponding control subjects. The healthy participants were mainly recruited from the hospital staff or their relatives, or from among the patients’ friends. All patients and control subjects were nonsmokers. Both groups had performed regular physical training two to three times per week, at least for the latest month, representing an activity score of 6. The activity score ranges between 1 and 8, with 1 representing no activity, and 8 representing training and competition on an elite level. The training could consist of participation in aerobics, dance or body pump classes, playing tennis, ice hockey or soccer, running, or cycling. Weight and height were measured in all participants in relation to the tests. BMI was calculated. FEV<sub>1</sub> was measured (Jaeger MasterScreen Body; Erich Jaeger GmbH; Würzburg, Germany) and was compared to reference values.

The study was approved by the Ethics Committee at Göteborg University, and informed consent was obtained from all patients and subjects, and from the parents of the children. The experiments complied with the Helsinki Declaration and Swedish law.

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**Table 1—Clinical Data in 33 Patients With CF and 20 Healthy Control Subjects**

<table>
<thead>
<tr>
<th>Variables</th>
<th>CF (n = 16)</th>
<th>CG (n = 8)</th>
<th>CF (n = 17)</th>
<th>CG (n = 12)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, yr</td>
<td>23.6 (6.2)</td>
<td>26.9 (6.6)</td>
<td>25.0 (5.0)</td>
<td>26.7 (5.8)</td>
</tr>
<tr>
<td>Height, cm</td>
<td>165.5 (4.6)</td>
<td>164.9 (5.3)</td>
<td>179.1 (7.4)</td>
<td>187.2 (6.4)</td>
</tr>
<tr>
<td>Weight, kg</td>
<td>60.3 (7.0)</td>
<td>61.2 (7.5)</td>
<td>72.6 (7.3)</td>
<td>73.7 (4.9)</td>
</tr>
<tr>
<td>BMI, kg/m²</td>
<td>22 (2.2)</td>
<td>23 (2.4)</td>
<td>23 (3.0)</td>
<td>23 (2.2)</td>
</tr>
<tr>
<td>FEV&lt;sub&gt;1&lt;/sub&gt;, % predicted</td>
<td>94 (19.2) [60–124]†</td>
<td>114 (9.8) [100–132]</td>
<td>90 (23.2) [41–126]‡</td>
<td>107 (9.9) [92–123]</td>
</tr>
</tbody>
</table>

*Values given as mean (SD) [range].
†p = 0.01 (compared to healthy control subjects).
‡p < 0.05 (compared to healthy control subjects).
Test Procedure

All subjects were tested at a research laboratory by a physical therapist who was accustomed to the testing procedure (E.M.T.) but was unaware of whether subjects were patients or control subjects. None of the patients or control subjects had been tested before. The procedure was equal for all participants. The standard instructions were given, and the subjects were allowed to familiarize themselves with each specific test. The tests were always performed in the same order as presented here. No recovery time was given except for time to move between the tests and the time for instructions. Verbal encouragement was not used during the specific tests, but instructions to undertake all of the tests with a maximum of effort were given in advance. All subjects wore a t-shirt, shorts, and fitness shoes. The subjects warmed up on a cycle ergometer for 5 min at 100 W of resistance and did 20 submaximal push-ups against a wall. The total time for the tests was 1 h and 15 min for all subjects. The following test protocol was used for all subjects.

Vertical jump (ie, counter movement jump) was performed on a jumping mat connected to an electronic timer (Time-it; Eleiko Sport; Halmstad, Sweden). The subject started in the upright position, and the jump was performed with both legs. Free arm swings were allowed. The movement started with bending the knees and was directly followed by an upward movement, ending with a maximal vertical jump and landing on both feet. Submaximal trials were allowed before the test started. The highest height (in centimeters) of three maximal tests was registered.

Hand-grip strength was measured with a portable instrument with a grip device and arm support (Grippit; AB Detector; Göteborg, Sweden) that enabled standardized arm and grip positioning. The grip device was connected to an electronic unit with an adapter for connection to a power supply. The grip device allowed the fingers and palm to be completely clasped around it. The force exerted against the transducer located in the grip device was displayed and recorded. Grip force (measured in newtons) registered the maximal voluntary contraction (MVC), and the mean value during 10 s registered the sustained MVC (SMVC). Both the right and left hands were tested, always beginning with the right hand. The test was performed three times with a 1-min rest between trials. The highest value was recorded.

Abdominal muscle strength was measured with sit-ups. The subject lay supine on a mat, hands together behind the neck, knees bent at 90°, and feet flat on the mat and 10 cm apart. The test leader held on to the feet. A sit-up was correctly performed when the subject bent the knees at 90°, and feet flat on the mat and 10 cm apart. The test was performed three times with a 1-min rest between trials. The highest value was recorded.

Arm/shoulder muscle strength was measured with push-ups. The subject was in the prone position on a mat, the hands placed at shoulder width under the shoulder joint, fingers forward, arms straight, head and body aligned, and toes on the mat 10 cm apart. The push-up was correctly performed when the subject bent the elbows to 90°, holding the body straight, and then straightened the elbows again. The subject was instructed to perform the test as fast as possible. When exhaustion occurred or if two incorrect push-ups were performed, the test was interrupted. The total number of push-ups was recorded.

Quadriceps muscle strength was measured with a hydraulically driven and microcomputer-controlled device (Kinetic Communicator II; Chattanooga Group; Chattanooga, TN), which operates in an isokinetic or isometric mode. The subject was positioned with a hip angle of 120° in the test chair. The axis of the knee joint was approximated to the axis of rotation of the dynamometer. The subject was stabilized with a strap around the waist. The leg-stabilization restraint was placed as far distally on the tibia as possible, while allowing full dorsal flexion of the ankle. The arms were crossed in front of the chest. In order to become accustomed to the test, the subject performed three submaximal concentric muscle actions of the knee joint. Three maximal concentric muscle actions were performed with 30 s of rest between the trials, at both 60° and 180°/s. Three maximal voluntary isometric contractions in knee extension were performed at a knee joint angle of 60°. The highest value was recorded. Both legs were tested. The starting leg was randomized. The force was measured in newton meters or newtons. All measurements were performed with applied gravity compensation.

A functional knee bend test was created to measure leg muscle endurance. The subject stood on the floor with the hips bent at 90°, a straight back and neck, feet 10 cm apart, and arms hanging. In that position, the level of the distal part of the middle finger was marked on the leg. The distance between the lateral malleolus and the lateral knee joint was measured with a tape measure, and half of that distance was marked. The subject was instructed to do as many knee bends as possible, touching the two marks, while keeping the back straight. The velocity was standardized with a metronome set at 120 beats/min, corresponding to 1 Hz. The test was performed until the subject was exhausted and was interrupted if the standardized procedure could not be sustained. The total number of knee bends was recorded.

Statistical Analysis

The conventional formula was used for calculations of mean (SD). The Student t test was used for comparisons between groups. The strength of correlation between the variables was assessed by using the Spearman rank correlation test. Statistical significance was set at p < 0.05 (StatView for Windows, version Xp; SAS Institute; Cary, NC).

Results

The patients with CF had normal anthropometric data, compared to age-matched and sex-matched control subjects (Table 1). Our study was representative for 90% of young CF patients in Göteborg, since those who declined to participate in the study did not differ from those in the study group when it came to anthropometric data or lung function. Ten percent of the patients were excluded from the study due to severe lung disease or pregnancy. The median physical activity level in the studied groups was 6,20 indicating that the basic physical activity level of our CF patients was similar to that of the control subjects. Despite good lung function, the FEV1 was significantly lower than that in control subjects for both men (p = 0.02) and women (p = 0.01) [Table 1], but did not differ according to gender among CF patients.

The results of the muscular strength and function tests are depicted in Table 2. The men with CF, but
The number of sit-ups performed in 30 s was the only parameter that correlated with FEV1 (p < 0.05) for men with CF. No gender differences could be seen in the total number of sit-ups between CF patients and control subjects. Hand-grip strength in both the right and left hands was significantly decreased in the women with CF, compared to healthy women. The isokinetic quadriceps muscle strength (180°/s) was significantly decreased in men with CF (p < 0.02), but neither gender differed from control subjects in the same test (60°/s). The women with CF did not accomplish the same number of knee bends as control subjects (p < 0.02). One male patient with CF was not able to perform the knee-bend test, and one female control subject did not perform the isometric quadriceps muscle test on the right leg because of knee pain.

Women had low arm-shoulder strength, and seven subjects with CF and two control subjects did not manage to perform any push-ups.

The mean values in all tests were lower for the CF patients than for the control subjects, except for the quadriceps isometric test in the right leg for men with CF. The significant differences were not associated with PI (n = 24) or PS (n = 9) [Table 3].

Discussion

This study showed good physical performance in terms of muscular strength and function, which was analyzed comprehensively in a young adult Swedish CF cohort of men and women with normal anthropometry. We found favorable physical status in the patients with CF, with basic physical activity scores similar to that of the age-matched and sex-matched control subjects, despite the existence of the complications normally found in this age group (ie, five patients had CF-related diabetes mellitus and seven patients had liver disease). Most of the patients also had severe cystic fibrosis transmembrane conductance regulator mutations and had received a diagnosis in early childhood. Thus, they were representative of most young adult patients at our center, and we found only slight impairment in muscular function, which was more prominent in the women.

Although lung function was significantly decreased, compared to control subjects, it was good for this age group6,8,11 and could not be related to muscular impairment since only sit-ups performed in 30 s in men with CF correlated with FEV1. Pancreatic status was not related to impaired muscular strength or function.
Previously, Lands et al. demonstrated correlation between leg strength, measured with an isokinetic cycle ergometer (expressed in watts), and lean body mass in CF patients and control subjects. We found a similar correlation between isokinetic quadriceps strength, at both 60° and 180°/s (data not shown), and lean body mass in CF patients (Fig 1). Unfortunately, we could not perform dual x-ray absorptiometry in the control subjects. Leg muscle endurance is important in daily life. The knee-bend test is a test of muscle endurance, and was, in our opinion, more functional and in accord with activities of daily living and many sports than endurance testing on a cycle ergometer. Therefore, it was interesting that this test yielded lower values in women with CF, but this discrepancy cannot be explained by differing activities.

Hand-grip strength has been shown to be related to upper extremity strength. The strength in the upper thorax, in the abdominal muscles, as well as in the arms/shoulders are important for thoracic posture and for facilitating coughing in patients with lung disease. Thus, it is interesting to measure in CF patients. The sit-up and push-up tests are recommended in the guidelines of the American College of Sports Medicine for measuring abdominal muscle strength and upper extremity strength. Pinet et al. concluded that CF patients with FEV₁ < 60% predicted had thicker and stronger abdominal muscles than did control subjects, and that this might be due to the heavier respiratory work performed by these patients. That interpretation was probably not valid in our patient population in which FEV₁ was 90% predicted and 94% predicted, respectively, in men and women. Intact lung function entails less heavy respiratory work. In the study by Pinet et al., the quadriceps isokinetic peak torque at 60°/s in the dominant leg, measured with a dynamometer (Cybex; Medway, MA), was 36% lower in the CF group, including both men and women, than in the control subjects. Our patients had 10% lower values, which

Table 3—Differences in Muscular Strength and Function Between Patients With CF With and Without Preserved Pancreatic Function Compared to Healthy Age-Matched and Sex-Matched Control Subjects*

<table>
<thead>
<tr>
<th>Variables</th>
<th>Women (n = 16)</th>
<th>p Value</th>
<th>Men (n = 17)</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right MVC, newtons</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CG (n = 8)</td>
<td>−45.5</td>
<td>0.02†</td>
<td>−25.6</td>
<td>0.41</td>
</tr>
<tr>
<td>PS (n = 5)</td>
<td>18.6</td>
<td>0.47</td>
<td>15.7</td>
<td>0.73</td>
</tr>
<tr>
<td>Right SMVC, newtons</td>
<td>−45</td>
<td>0.06</td>
<td>−30.6</td>
<td>0.26</td>
</tr>
<tr>
<td></td>
<td>23.9</td>
<td>0.41</td>
<td>18.1</td>
<td>0.64</td>
</tr>
<tr>
<td>Left MVC, newtons</td>
<td>−65.7</td>
<td>0.001‡</td>
<td>−21.4</td>
<td>0.43</td>
</tr>
<tr>
<td></td>
<td>−3.2</td>
<td>0.91</td>
<td>−3.7</td>
<td>0.93</td>
</tr>
<tr>
<td>Left SMVC, newtons</td>
<td>−61.7</td>
<td>0.002‡</td>
<td>−19.3</td>
<td>0.47</td>
</tr>
<tr>
<td></td>
<td>14.9</td>
<td>0.52</td>
<td>−8.9</td>
<td>0.82</td>
</tr>
<tr>
<td>Sit-ups in 30 s, No.</td>
<td>−2.1</td>
<td>0.12</td>
<td>−3.1</td>
<td>0.03‡</td>
</tr>
<tr>
<td></td>
<td>−0.6</td>
<td>0.72</td>
<td>−2.9</td>
<td>0.25</td>
</tr>
<tr>
<td>Sit-ups total, No.</td>
<td>−15.3</td>
<td>0.20</td>
<td>−9.6</td>
<td>0.15</td>
</tr>
<tr>
<td></td>
<td>5.3</td>
<td>0.51</td>
<td>−12.7</td>
<td>0.14</td>
</tr>
<tr>
<td>Push-ups, No.</td>
<td>−6.3</td>
<td>0.16</td>
<td>−5.0</td>
<td>0.10</td>
</tr>
<tr>
<td></td>
<td>0.8</td>
<td>0.87</td>
<td>−3.3</td>
<td>0.47</td>
</tr>
<tr>
<td>CMJ, cm</td>
<td>−2.1</td>
<td>0.35</td>
<td>−0.8</td>
<td>0.68</td>
</tr>
<tr>
<td></td>
<td>3.2</td>
<td>0.30</td>
<td>3.7</td>
<td>0.20</td>
</tr>
</tbody>
</table>

*md = mean difference; CMJ = counter movement jump.
†p < 0.05.
‡p < 0.01.

Figure 1. Isokinetic strength of the right leg (Nm [newton-meters]) at 60°/s in patients with CF (n = 28) in relation to lean body mass (r = 0.800; p < 0.0001). ◯, women; ●, men.
was not significantly different from control subjects. Interestingly, the quadriceps isokinetic peak torque at 180°/s in the left leg was significantly decreased in men, although the percentage decrease was the same. The results therefore suggest a slightly lower quadriceps function, thus confirming in our patients the results of Pinet et al., although our patients had better lung function. We could not confirm the previously shown correlation between lung function and quadriceps strength.

It is well-known that healthy women have 40 to 60% less muscular strength in the upper body and about 25% less muscular strength in the lower body compared to men. This was confirmed in our study (Table 2) and also was found to be relevant for CF patients. The men with CF differed in muscle strength and function in only two tests, compared to the male control subjects, but women differed in four tests, which were not the same tests as those in which the men differed. Elkin et al. reported that adults with CF had significantly less quadriceps and hand-grip strength than did control subjects. They did not differentiate according to gender, and it is thus not possible to compare their results with ours.

The differences between patients with CF and control subjects were probably not due to reduced muscle mass since the studied patients had normal lean body mass. The weakness could not be due to the use of corticosteroids, since they are rarely prescribed to our patients. Only one male patient had periodically received such therapy.

In conclusion, our study mainly showed similar muscular strength and function in young adult patients with CF when compared to age-matched healthy control subjects with similar basic physical activity. Slight muscular weakness was found in women with CF, which was indicated by lower endurance in hand and leg muscles, and, in men with CF was indicated by lower endurance in abdominal muscles and quadriceps. The results confirm the presence of impaired endurance even in well-trained patients but would hardly have been disclosed if the gender perspective had been omitted when analyzing the control subjects. The issue of whether the impairment in muscular strength and function are due to metabolic, hormonal, or neuromuscular factors requires further investigation.

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