Evaluation of aerobic exercise capacity and daily functioning of patients with cystic fibrosis

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Introduction: Exercise capacity of CF patients is used as a parameter of disease status and progress. In Israel, there is no data on exercise capacity and daily function levels of CF patients.

Objectives: The target of this study were collecting data on CF patients’ aerobic capacity, validating a physical activity questionnaire in Hebrew and identifying exercise limits. Results will provide a basis for building individual home exercise programs to help stabilize patients’ clinical condition.

Methods: Patients (age ≥ 12 yrs) underwent cardiopulmonary exercise test, a six minute walk test and filled-in physical activity questionnaire. Correlations between test results were statistically analyzed.

Results: Out of 17 patients [8 males, 9 females, mean age: 20.3 ± 6.4 years (range: 12–34)] who completed the study, Pulmonary function test revealed mean FEV1% of 87 ± 10.5. Exercise test showed mean “6 minutes walk” test distance (m) of 629.9 ± 41.0. Mean VO2 max (ml/min/kg) for males: 46.3 ± 10.0. Mean VO2 Max (ml/min/kg) and for females: 39.4 ± 5.7. According to the questionnaires mean time per week of physical activity was 300 min for males and 120 min for females. Pearson correlation parameter of r = –0.74 was seen between Borg scoring and VO2-max in cardiopulmonary stress test.

Conclusion: Although male patients engaged with higher levels of physical activity compared to female patients, the VO2 max values of male patients correlated only with moderate aerobic capacity level. In addition to cardiopulmonary exercise test we found that a questionnaire is an effective tool to assess daily physical activity in CF patients. More patients are needed to assess correlations between test results.

Improvement of body composition parameters after an individualized training program in young patients with CF

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Background: Despite significant improvements in the CF treatment, alteration in body composition is still a common problem which affects the quality of life.

Objectives: This study try to improve the elements of body composition and to observe the changes after 6 months of supervised physical individualized training on patients with CF.

Methods: We conducted a 6 months prospective study, on 20 patients (between 14–18 years), in the Romanian National C.F. Centre. The evaluation consisted in: analysis of corporeal composition through bioimpedancy (In-Body 720 device). In order to rich our purpose regarding the body composition the subjects benefit by a dietary recommendations and individualized training program 4 times/week with heart rate between 65–80%.

Results: We noticed important changes in Weight (from 44.8±16.06 to 46.23±16.38, p = 0.0023), proteins (from 7.52±2.422 to 8.425±2.494, p = 0.0002) and Skeletal muscle mass (20.23±7.226 to 22.03±7.277, p = 0.0002), and also small changes regarding body water (Intra cellular water increased from 17.08±5.554 to 17.82±6.594, and Extra cellular water from 10.60±3.274 to 11.47±3.478).

Conclusion: At the end of the study we observed a positive evolution of the body composition for the majority of patients included in the study. Impaired breathing and bacterial infections trigger the body to burn more calories than usual and it is necessary the increasing of lung function through physiotherapy and dietary supplement.

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Tolerance of nebuliser challenge in children less than a year of age

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Background: Nebuliser therapy is increasingly utilised in infants with Cystic Fibrosis, including colistin or tobramycin for the treatment of Pseudomonas aeruginosa infection, and DNAse or hypertonic saline (HS) as adjuncts for secretion clearance. There may however be concern over the tolerability of nebulised therapy, particularly HS, in patients less than 1 year of age. Prior to commencing nebuliser therapy we perform a formal treatment challenge under physiotherapist supervision. We reviewed treatment challenge in infants less than a year of age compared to older children.

Methods: Prospective evaluation of formal nebuliser challenge in all newly initiated nebuliser therapy.

Results: Over a 15 month period 37 nebuliser challenges were performed in 27 children – 21 to 7% HS, 1 to 3.5% HS (previously unable to tolerate 7% HS), 6 to Tobramycin, 5 to Colistin and 4 to DNAse. There were 5 challenge failures – 2 to tobramycin via eflow nebuliser (both subjects subsequently tolerated with medix nebulisers), and 3 to HS – one subject had >10% decline in FEV1 after 3.5% HS; one age 2 years vomited after HS challenge, and one age 9 months would not tolerate the face mask. There was no significant difference in tolerability between infants and older children – in 6 challenges less than 1 year of age (5 for 7% HS and 1 for Colistin) only one was not tolerated due to the mask (17%), compared to 4 failures in older children (13%). All patients who tolerated their nebuliser challenge were successfully commenced on regular therapy.

Conclusions: Nebuliser challenges are feasible and well tolerated even in infants less than 1 year of age, and are predictive of therapy tolerance.

Improvement of body composition parameters after an individualized training program in young patients with CF

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The I-neb Adaptive Aerosol Delivery (AAD) System is designed to analyze patient breathing and emit aerosol only during the inspiratory phase. The I-neb AAD System can be operated in tidal breathing mode (TBM), in which the user breathes in a normal manner during treatment, or target inhalation mode (TIM), in which the user is guided to longer inhalations via feedback from the I-neb device, depending upon patient preference. Each I-neb device is equipped with a patient logging system (PLS) that records details about each treatment taken via the device, including treatment time and mean inhalation time per breath per treatment. We analyzed PLS data from a 13-week handling study conducted in 49 adult patients with cystic fibrosis to determine the association of mean inhalation time (time spent inhaling during each breath) per treatment with the time taken to complete a treatment (treatment time). Data for 43 patients was analyzed in terms of inhalation time per breath and treatment time.

Table 1. Cumulative number of patients and mean treatment times for increasing inhalation time.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Mean inhalation time category (s)</th>
<th>TBM</th>
<th>TIM</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Number of patients (n)</td>
<td>Mean treatment time (min)</td>
<td>Number of patients (n)</td>
</tr>
<tr>
<td>&gt;0.5</td>
<td>20</td>
<td>5.1</td>
<td>23</td>
</tr>
<tr>
<td>&gt;2</td>
<td>12</td>
<td>3.9</td>
<td>22</td>
</tr>
<tr>
<td>&gt;3</td>
<td>10</td>
<td>3.7</td>
<td>20</td>
</tr>
<tr>
<td>&gt;5</td>
<td>5</td>
<td>3.5</td>
<td>17</td>
</tr>
<tr>
<td>&gt;8</td>
<td>2</td>
<td>4.1</td>
<td>5</td>
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</table>

Most patients using TIM had inhalation times over 5 s, whereas most TBM patients had inhalation times under 5 s. The mean treatment time for patients using TIM was between 1 and 2 minutes shorter than for patients using TBM. These results suggest that patients using TIM would have shorter treatment times.