Cystic Fibrosis and Physical Activity
Total Energy Expenditure and Physical Activity Levels in Children and Adolescents with Cystic Fibrosis

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Preface
We would like to thank Afsaneh Roshanai and Roger Olsson for support, data and help with content suggestions. We also would like to thank Carina Ahlstedt for helping us with complex SPSS that made our days less enjoyable.

Thank you again.

Sincerely,
Tova Dahne and Tatyana Filonova.
Uppsala, December 2011
Abstract
Aim: The aim with the current study was to examine the physical activity levels (PAL) in a group of children and adolescents with cystic fibrosis (CF) and compare PAL-values between boys and girls. Further, the aim was to look at the total energy expenditure (TEE) estimated with an activity diary and measured with the accelerometer and compare values between these two measurement methods. Method: The sample consisted of 29 children and adolescents diagnosed with CF where PAL and TEE was measured during a three-day registration with activity diary and the ActiCal© accelerometer. The data was analyzed with the Mann-Whitney U-test. Results: The results indicated that the girls had a slightly higher median PAL-value than the boys. In addition, the range between the lowest and highest PAL-value was larger among the boys. The lowest PAL-value was lower among the boys than among the girls and the highest value was similar to the girls’ highest PAL-value. Overall, the subjective estimation of the PAL-values were slightly higher than what was objectively measured by the accelerometer. There was a slight difference between TEE estimated with the activity diary compared to the TEE measured by the accelerometer. There was no significant difference in PAL-values between boys and girls. Conclusion: There was no significant difference in PAL-value between boys and girls and that there was a slight difference in TEE estimated with the activity diary and measured with the accelerometer. Both methods of data collection are reliable enough when used together to produce a valid estimation of CF-children’s activity levels and TEE. The fact that both PAL and TEE values were higher in the activity diary may be because the participants perceived a higher level of physical activity then what was objectively measured with the accelerometer.

Keywords: Cystic fibrosis, children, adolescents, energy expenditure, activity diary, accelerometer, ActiCal©.

Abbreviations

<table>
<thead>
<tr>
<th>Abbreviation</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>BMR</td>
<td>basic metabolic rate</td>
</tr>
<tr>
<td>EE</td>
<td>energy expenditure</td>
</tr>
<tr>
<td>TEE</td>
<td>total energy expenditure</td>
</tr>
<tr>
<td>PAL</td>
<td>physical activity level</td>
</tr>
<tr>
<td>AD</td>
<td>Activity diary</td>
</tr>
<tr>
<td>AC</td>
<td>Accelerometer</td>
</tr>
</tbody>
</table>
Sammanfattning


Nyckelord: cystisk fibros, barn, ungdomar, energiförbrukning, aktivitetsdagbok, accelerometer, ActiCal©
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1. Introduction

1.1 Cystic fibrosis

1.1.1 Physiology of cystic fibrosis
Cystic fibrosis is caused by mutations in the CF transmembrane conductance regulator (CFTR) protein and leads to abnormal chloride transport resulting in viscous secretion in the pulmonary, gastrointestinal, endocrine and reproductive system and an increasing salt loss in sweat (Philpott, Houghton & Luke, 2010).

CF causes damage in several organs and especially affects lungs, causing repeated lung infections, and pancreatic insufficiency (see Appendix 3). This leads to a reduced volume of pancreatic secretion of HCO$_3^-$ and malabsorption in the gastrointestinal system. Mucus becomes more viscous and the risk of obstruction of pancreatic ducts increases. Clogged ducts can quickly result in local inflammation. Islets of Langerhans become damaged only after a few years resulting in CF-related diabetes, which is a combination of type-1 and type-2 diabetes. A side effect of lung infections is reduced appetite and vomiting which increases the malnutrition, leading to exacerbation of the lung infection, which in turn causes a difficult cycle of malnutrition (Ratjen & Döring, 2003).

1.1.2 Treatment
At present there is no single pharmacological treatment for cystic fibrosis thus it is treated only symptomatically. Getting rid of excessive mucus is considered to be one of the most important goals of CF treatment. This can be done by inhalation therapy, where a person is inhaling bronco-dilators or hypertonic saline solution, which hydrates the viscous mucus. Chest physiotherapy is a good non-pharmacological treatment that not only decreases obstructivity, but also helps build strength, condition and also maintain flexibility of the lungs (Eriksson et al, 2009).

It is vital to maintain the nutrition of a CF-patient, since it positively correlates with the overall health status. Intake of nutritious drinks and supplements should be considered if the patient has
trouble maintaining the weight. In the most severe cases one should consider the usage of a sonde.

If exacerbation occurs it is important to set in a course of antibiotics, even if the infection proved to be viral. Careful information and demonstration of how to administer drugs via intravenous catheter will make it possible for the patients to self-administer the antibiotics without it interrupting their daily lives. There has also been attempts to control the constant inflammation that arises in the airways due to the bacteria colonizations. According to many studies, some drug inhalations were useful for people in certain age group, but not others. Other inhalation drugs had a good effect on lung function, but had an unwanted side effect. In other words, the search for the most optimal drug therapy with the least side effects is still on-going. Lung transplantation is also raised as the alternative treatment for cystic fibrosis. However, the survival rate has proven to be lower than in other transplantation and thus should be taken as the last resort (Rajten & Döring, 2003).

1.1.3 Prevalence
The prevalence of CF in Sweden is currently 600 individuals and about 20 children are born with the disease each year (Socialstyrelsen, 2005a). According to the United States Registry, the median survival age rose from 14 years in 1998 to 32 years in the year 2000, due to early detection, different treatments in specialized centers and socio-economic status (Ratjen & Döring, 2003).

1.1.4 CF in healthcare
Since CF is a multi faceted disease it is important to see the overall picture regarding the patient and his/her family. Physicians alone cannot provide adequate care; a team consisting of nurses, nutritionists, respiratory therapists and social workers is necessary to achieve the best outcome (O’Sullivan & Freedman, 2009). Sweden has four healthcare centers that are specialized in CF-care. In the CF center, the patient meets a team consisting of doctors, nurses, physiotherapists and dietitians, as well as counselors and psychologists (Socialstyrelsen, 2005a).
1.1.5 Nutrition in cystic fibrosis
Pancreatic deficiency makes the absorption of fat impaired. This can be compensated by pancreatic enzyme replacement therapy (PERT). This means swallowing capsules containing digestive enzymes with every food intake, but the dosages differ greatly between different hospital standards. At the same time, it is also important to ensure that the patients are digesting large amounts of fat-soluble vitamins (A, D, E, K), to compensate for the lack of reserves (Eriksson et al, 2009). For individuals of two years of age and older it is recommended to have energy intakes that are greater than the standard for the population. Research results have proved that energy intakes ranging from 110% to 200% of the standard for the population, not only help maintain weight in adults but also help children gain weight at an age-appropriate rate (Stallings et al, 2008). Pursuant to Ratjen and Döring (2003), those who have a deficient nutritional status have a higher risk of getting chest infections compared to those who have a good nutritional status. Needless to say, it is of great importance to follow the child’s nutritional status closely to avoid exacerbation and further complications.

1.1.6 Physical activity in cystic fibrosis
Physical activity has become a broadly accepted and important part of the rehabilitation programs in CF-care. Several beneficial effects have been reported, and studies shows that exercise training improves cardiopulmonary fitness (CPF) in children and adolescents with CF. Furthermore, Williams, Benden and Stevens (2010) report that physical activity may ameliorate aerobic and anaerobic capacity, strengthen ventilation muscles and helps clear airway sputum. Possible risk factors can be hypoxia due to respiratory insufficiency and severe sodium losses because of excessive salt excretion when sweating.

The result of Williams et al (2010) is strengthened in the article by Philpott, Houghton and Luke (2010) where the authors claim that balanced and individually adapted physical activity may slow down deterioration in lung function in children with CF and increase survival rates. Aerobic fitness such as swimming, jogging and walking can improve endurance of respiratory muscles while strength training can improve forced expiratory volume (FEV₁). The above-mentioned research group also present potential risks with physical exercise in children with CF and
highlights the degree of the lung disease and subnormal ventilatory capacity as the major limitations. Bronchial narrowing, bronchospasm, mucous plugging and reduced alveolar ventilation may be the causes of these limitations. Resting energy expenditure is 5% to 25% higher in CF children, complicating physical exercise further. Aggravating conditions such as greater salt loss in sweat and developed diabetes mellitus demand good control over food and fluid intake because of the risk of hyponatremic dehydration and hypoglycemia. CF children with liver dysfunction or splenomegaly also have a higher risk of organ damage while participating in contact or collision sports (Philpott et al. 2010). One should also note the gender differences regarding the activity levels when children reach puberty. At that period, boys, compared to girls, have a much higher level of activity (Williams et al, 2010).

In a study conducted by Gruber, Orenstein, Braumann, Paul and Hüls (2011) where they investigated the response of an in-patient exercise program in children with CF and compared the results between boys and girls, showed improvement in fitness levels among both girls and boys. A small but significant sex-based difference in results regarding physical activity performance was noted and authors suggests this difference could be ascribed to physiological and morphological differences between the sexes.

1.1.7 Recommendations regarding physical activity in children

According to the Nordic Nutrition Recommendations (NNR) physical activity is any bodily movement that is made by contraction of muscles that increases energy expenditure. The recommended minimum physical activity for children and adolescents according to the NNR is 60 minutes a day of moderate to vigorous activity. It is a necessity for the child’s normal growth and development of cardio-respiratory endurance, muscle strength, flexibility, motor skills and agility. Physical activity is also associated with psychological well-being and has a strengthening effect on self-esteem. NNR recommends a PAL value at 1.8 for adults to maintain good health. There are only recommendations regarding PAL when it comes to adults but two tables with different activity levels and ages are represented below (Livsmedelsverket, 2005).
Table 1.1. PAL: Girls 10-13 years (14-17 years)

<table>
<thead>
<tr>
<th>Low physical activity</th>
<th>Moderate physical activity</th>
<th>High physical activity</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.50 (1,45)</td>
<td>1.70 (1,65)</td>
<td>1.90 (1,85)</td>
</tr>
</tbody>
</table>

Table 1.2. PAL: Boys 10-13 years (14-17 years)

<table>
<thead>
<tr>
<th>Low physical activity</th>
<th>Moderate physical activity</th>
<th>High physical activity</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.55 (1,60)</td>
<td>1.75 (1,80)</td>
<td>1.95 (2,05)</td>
</tr>
</tbody>
</table>

(Nordic Nutrition Recommendations, National Food Administration, 2004)

When it comes to difference in PAL between healthy boys and girls, researchers Hoos and co-workers (2003), who studied physical activity levels in children and adolescents, claim that there was no significant difference between boys and girls.

1.1.8 Measurement methods of physical activity and energy expenditure

Methods to measure physical activity can be divided into two groups, objective and subjective methods. The most frequently used objective methods are direct observations, pedometers, accelerometers and heart rate monitors. In direct observation, a trained individual observes the direct recording of physical activity levels. This is one of the first methods used to assess physical activity and is considered to be a reliable one. The most common subjective method to measure physical activity is self-reporting. The method is more practical when it comes to finding a good method to use in practice, but the validity of individual measurement has room for errors when it relies on a person’s willingness to record daily and accurately. The self-reporting can include both daily diaries on paper or electronic formats and recall questionnaires (Reiser & Schlenk, 2009). For measuring energy expenditure, the method of doubly labelled water (DLW) is recognized as the “golden standard” and is a measuring method which other methods are validated against. It can give information about a person’s TEE over a period of 4 to 20 days and begins with the test person drinking a little amount of water containing a certain amount of stable, non-radioactive isotopes of hydrogen and oxygen. The rate of which the isotopes exit the body can then be measured through urine, saliva or blood. By estimating or
measuring the respiratory quotient one can then calculate the oxygen uptake for the test period and thus know the energy expenditure of a test person (Bellardini, Henriksson & Tonkonogi, 2009).

When it comes to describing physical activity one well established method of calculating a person’s daily physical activity is PAL, which is the result of dividing total energy expenditure (TEE) and basic metabolic rate (BMR). TEE is the sum of an individual’s basic metabolism, physical activity and the food’s thermogenic effect. For children’s and adolescents’ growth is also taken into account when calculating total energy need. BMR is the energy consumption an individual has in one day, when awake and in physical and psychological rest (Andresson & Göranson, 2006).

1.1.9 Nurse’s role

CF is a demanding disease both for the patients and their families and requires extensive involvement from many different healthcare professionals. According to the Competence Description for Registered Nurses (Socialstyrelsen, 2005b), nurses have responsibility regarding organizing and/or participating in teamwork around the patient and shall ensure that the patients, relatives and close ones get adequate information, education, support and guidance so that they can participate in care and treatment.

Dorothea Orem’s definitions and nursing theories can be used to explain nurse’s role in the treatment of cystic fibrosis. She defines self-care as a practice of activities that the people initiate in order to maintain their well-being. When an individual for some reason cannot maintain its self-care, therapeutic self-care demand arise. Therapeutic self-care demand is the totality of the care activities needed to meet a person’s requirement for self-care, which in turn culminates in self-care requisites, which are reasons for self-care activities and can be divided into three groups: developmental, universal and those associated with health deviation. The latter ones are related to changes in a body’s function and/or structure, to which children with cystic fibrosis can be said to belong. In Orem’s Theory of Nursing Systems she outlines a nurse’s three systems, through which she can help a patient fulfill its self-care requisites: wholly compensatory
system, partly compensatory system and supportive-educative system. When treating cystic fibrosis in children a supportive-educative system is perhaps the optimal one to use. Since self-care is vital in the treatment of cystic fibrosis, it is important for the nurse to regulate the exercise and to further help develop a person’s capabilities in order to overcome the struggle with the disease (Coldwell Foster, 2010).

1.2 Formulation of problem
According to the literature, cystic fibrosis seems to be a complex disease with no single treatment. One important aspect in treatment of cystic fibrosis is physical activity. Without a doubt there will be times when nurses might come across caring for children with cystic fibrosis or patients with other similar genetic disorders. Therefore, it is important to learn about these patients and about which measurement methods that are used in CF-research today and if there are any gender differences in physical activity levels.

1.3 Aim
The aim of this paper was to examine the TEE-values between two measuring methods, accelerometer and activity diary, on a sample of children and adolescents at a CF-center at Uppsala University Children’s Hospital. Further the aim was to compare the PAL-values between these boys and girls in the sample.

1.4 Specific study questions
1. Are there any differences between girls’ and boys’ physical activity level (PAL)?
2. Were there any difference in values regarding the children’s and adolescents’ total energy expenditure (TEE) between the activity diary and the accelerometer?
2. Method

2.1 Design

The study is descriptive and quantitative and is based on the data from an already performed study.

Sample
The sample consisted of 29 children and adolescents diagnosed with cystic fibrosis between the ages of 5 to 17 who came to the Center for Cystic Fibrosis at the Uppsala University Children’s Hospital.

Inclusion criteria
Children between 5 and 17 years of age diagnosed with cystic fibrosis.

Exclusion criteria
Children with multiple illnesses, those who had exacerbation during the study, or who did not have fully completed activity diaries.

2.2 Data collection method and procedure

With a sample of 29 children, the researchers made fundamental measures of each child’s body and foisted them with an activity diary and an apparatus that measures the body’s activity level (an ActiCal©) both on a wrist and ankle. In the activity diary the children were asked to estimate their activity levels ranging from one to nine, with one meaning sleep and nine - excessive training, four times each hour during three consecutive days. When estimating six or higher in a diary, they were asked to write down what they were doing at that time point. After three days the activity diaries and accelerometers were returned to the researchers to gather the data for analysis. The activity diary was based on one method originally described by Bouchard, Tremblay, Leblanc, Lortie, Savard and Theriault in 1983, which was further developed by Bratteby, Sandhagen, Fan and Samuelson in 1997 (Bratteby et al, 1997). This method was validated against doubly labeled water (DWL). The AktiCal© accelerometer was validated in a
study conducted by Puyau, Adolph, Vohra, Zakeri and Butte (2004), where they compared the ActiCal© accelerometer with a continuous four hours lasting measurements of energy expenditure (EE) in a respiratory room calorimeter and one hour long measurements in an exercise laboratory using a portable calorimeter and treadmill on 32 children, aged 7 to 18 years.

2.3 Data analysis

Data from the children’s and adolescents’ activity diaries and measurements from the accelerometers were available at the Center of Clinical Nutrition and Metabolism at Uppsala University Children’s Hospital and was structured and presented in an Excel file. Participants’ initials and year of birth were used as code to preserve the confidentiality. TEE was calculated with data from the activity diary and basic metabolic rate (BMR) was calculated according to the Schofield equation, which takes sex, age and weight in consideration (Andersson & Göranzon, 2006). Values that were relevant for the study questions were age, sex, mean value of TEE measured over three days with activity diary and accelerometer (kcal/d), and mean value of PAL measured over three days with activity diary and accelerometer (PAL/d). The data was analyzed with Statistical Package for Social Sciences (SPSS, v. 20.0.0). Kcal was the chosen unit since it is the most common unit used when calculating energy in food in daily life.

During the analysis, it has been noted that some subjects were either over or under the age group selected or had some missing data, which is why they were omitted from the study. Unfortunately, this resulted in a large difference in number of boys (n=19) and girls (n=10). The selected data was analyzed with the Mann-Whitney U-test which is the test recommended when the sample is small (Ejlertsson, 2003). According to Polit and Beck (2007) it is advisable to delete subject’s data when a significant amount of data is missing though the sample is small. The significance level was set at $p < 0.05$. 
2.4. Ethical considerations

The study is based on a research conducted by a large medical research group, which has already received approval from the Regional Ethical Review Board on 28th of November 2003 (reg. no. 03-414) and informed consent was obtained from the participants’ parents. The collected data was treated confidentially and information about the subjects was kept at the Center of Clinical Nutrition and Metabolism at Uppsala University Children’s Hospital. Authors had approval from the supervisor and head researcher to use information from the larger study. Requirements that are needed for doing the study, such as information about the study (see Appendix 3), a right to withdraw at anytime, confidential treatment of personal information and, ultimately, the usage of data only for research purposes were met (CODEX, 2011).

One ethical dilemma that occurs naturally is that when sexually active older boys and girls are asked to meticulously write down every activity that they do, they might, even unconsciously, perceive this as an invasion of privacy, which in turn might affect their compliance with the study.

3. Results

3.1 Differences between boys and girls regarding PAL measured with activity diary and accelerometer during three days

PAL values were slightly higher among the girls in comparison to the boys and the standard deviation was higher in the boys group (Table 1.3). However, there was no significant difference in the median value. The score range was wider in boys compared to the girls and the boys’ lowest value were lower than in the girl group (Table 1.3).
Table 1.3. Results for PAL value measured with accelerometer

<table>
<thead>
<tr>
<th></th>
<th>N</th>
<th>Mean</th>
<th>SD</th>
<th>Median</th>
<th>Range</th>
<th>IQR</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Boys</td>
<td>17</td>
<td>1.53</td>
<td>0.43</td>
<td>1.62</td>
<td>1.30 - 1.75</td>
<td>0.30</td>
<td>&gt; 0.05</td>
</tr>
<tr>
<td>Girls</td>
<td>8</td>
<td>1.62</td>
<td>0.11</td>
<td>1.58</td>
<td>1.52 - 1.72</td>
<td>0.22</td>
<td>&gt; 0.05</td>
</tr>
</tbody>
</table>

The results indicated a moderate difference between girls’ and boys’ mean values (Table 1.4). The low difference in standard deviation means that the spread of the values is small. Even here the range was bigger than among the girls. There was also a difference in mean and median values of the activity diary and accelerometer measurements in the girls group.

Table 1.4. Results for PAL value measured with activity diary

<table>
<thead>
<tr>
<th></th>
<th>N</th>
<th>Mean</th>
<th>SD</th>
<th>Median</th>
<th>Range</th>
<th>IQR</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Boys</td>
<td>19</td>
<td>1.69</td>
<td>0.51</td>
<td>1.60</td>
<td>1.42 - 1.95</td>
<td>0.36</td>
<td>&gt; 0.05</td>
</tr>
<tr>
<td>Girls</td>
<td>10</td>
<td>1.79</td>
<td>0.20</td>
<td>1.77</td>
<td>1.61 - 1.96</td>
<td>0.26</td>
<td>&gt; 0.05</td>
</tr>
</tbody>
</table>

3.2 Differences in TEE between activity diary and accelerometer

Table 1.5 Results for TEE measured with activity diary and accelerometer.

<table>
<thead>
<tr>
<th></th>
<th>N=</th>
<th>Mean TEE (kcal)</th>
<th>SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Activity diary</td>
<td>29</td>
<td>2404,7</td>
<td>689,7</td>
</tr>
<tr>
<td>Accelerometer</td>
<td>25</td>
<td>2080,6</td>
<td>727,6</td>
</tr>
</tbody>
</table>

There was a minor difference between the mean TEE values from the activity diary compared to the accelerometer values. The number of subjects having the accelerometer and the activity diary was similar.
4. Discussion

A slight difference between TEE estimated with activity diary compared to the TEE measured by the accelerometer was observed. The subjective estimation of the PAL-values was slightly higher than what was objectively measured by the accelerometer. Overall, there was no significant difference in PAL-values between boys and girls.

4.1 Result discussion

4.1.1 Difference in PAL value between boys and girls
According to the results there was a difference in the PAL-value between boys and girls estimated with the activity diary and measured by the accelerometer. Girls had a slightly higher level of PAL than boys. However, the girls group was significantly smaller than the boys group due to loss of data. This means that one cannot draw any valid conclusions regarding potential difference between boys and girls, because the result might as well depend on a chance. Therefore, the result was not significant. Though the difference in group sizes in the current study may have influenced the results, a comparison was made to the results of a review study by Hoos, Gerver, Kester and Westerterp (2003), where the authors collected data from 17 studies which compared PAL-values in boys and girls. According to the results of the review article no significant difference was found. So, it could be assumed that even with a larger sample and more equally distributed groups the non-significant results may have remained.

4.1.2. Differences in TEE between activity diary and accelerometer
The differences in TEE-values estimated by the activity diary and measured by the accelerometer were minor with TEE-values from activity diary being slightly higher than those by the accelerometer. This might be explained by that the children estimated their activity levels to be higher than what they actually were.

Authors speculate that one reason for why the CF children and adolescents reported a higher TEE after three-day measurement with an activity diary compared with TEE measured by the
accelerometer may be that the impact of the disease on respiration, making physical activity harder, which in turn may lead to that the perceived level of physical activity is higher than the objectively measured. The results of this study differ to that of the study made by Machado-Rodrigues et al (2011), where the researchers investigated the agreement between the values of the accelerometer and the activity diary in adolescents during three days. In their paper the adolescents seem to underestimate activity levels compared with accelerometer values. On the other hand, the research was done on mostly overweight adolescents, which suggests that the participants’ perceptions differ from the ones that CF-children have, due to the explanation above. Overall the current sample was small, but on the other hand the prevalence of CF in Sweden is low and the number of children and adolescents who have the disease is also small.

4.2 Method discussion

When conducting the analysis more of the girls’ data had to be removed due to missing bits of data. That made the number of people in girl-group lower than the number of people in the boys group, which may have skewed the result. In future studies though, to handle the problem with missing data, one can use imputation, which involves calculating an estimate of the missing value and replacing it with the estimate, tentatively by using mean substitution or median substitution (Polit & Beck, 2008). Since the number of subjects having accelerometer and activity diary was similar, one can say that the result was reliable due to rather equal distribution.

4.2.1 Activity diary

The activity diary is a more or less subjective measurement and is based on the child’s and adolescent’s own perception of the amount of time they have performed a certain activity and the degree of effort they experience during the activity. The activity diary is cost-effective and easy to use, characteristics that are often wanted in studies of this kind. The activity registration is done retrospectively and this may have led to over- or underestimations regarding the amount of time the activity took and how strenuous the activity was. Since the age limits in this study were 5-17 years some of the children were too small to fill out their activity diary by themselves, and a parent had to do it for them, leading to further risk for misregistrations. The activity registration
took place during three consecutive days and no account was taken regarding if these days were representative. The first day of registration was the day when the child or adolescent visited the CF-center at Uppsala University Children’s Hospital and the following days could be during the week or on the weekend, possibly influencing the child's/adolescents activity rate.

4.2.2. Accelerometer
The ActiCal© accelerometer was validated by Puyau et al. (2004) and is by the authors considered to be a valid way of measuring physical activity. The accelerometer is a good choice of equipment due to that it’s cheap compared to other measuring systems, suitable for all age groups and requires little effort from the participant. Furthermore, it is capable of objectively measuring the total physical activity, is validated and can detect a change in activity. On the other hand, when placed on only some body parts, it might not be able to measure all activities made by the participant. The data received can sometimes be difficult to interpret and process. Even if wearing the accelerometer requires very little effort from the participant, in some age groups, for instance, adolescents, compliance can still wane for different reasons (Medical Research Council, 2011). In this study, the activity registration by accelerometer took place during three consecutive days. In a review article written by Trost, McIver and Pate (2005), it was concluded that for the accelerometer to achieve reliability the number of monitoring days should be 4 to 9. The fact that the monitoring period in this study only was three days may have affected the result.

4.3 Clinical application and future research possibility

Orem’s Theory of Nursing Systems fits well during different phases of cystic fibrosis disease. When a patient fulfills its self-care well the nurse acts through a Supportive - Educative System, giving advice and helping develop a patient's self-care. When exacerbation occurs and the patient needs to be treated in the hospital, the nurse’s role is more likely to root from a Partly Compensatory System, where the nurse performs needed self-care measures and compensates for the patient’s limitations, while assisting him in resuming his control over the disease (Coldwell
Foster, 2010). Knowing both the medical and nursing approaches on treatment of CF, a nurse has an excellent opportunity to adapt the care to fit the patient’s need in whatever state he is in.

As seen in this study, CF is a complicated and multifaceted disease that demand individually adapted care from several different healthcare professionals. Though physical activity is widely accepted, but not a sufficiently researched part of CF-care, yet today, there is no standard when it comes to recommendations regarding physical activity in CF-care. Authors see that the childrens and adolescents with CF, as well as their families and the healthcare professionals who work with them, would benefit from having a more solid platform of recommendations to rely on when it comes to physical activity. Therefore, authors would like to see that the research regarding CF and physical activity continues and is focused on developing a platform of recommendations, making life easier for the children and adolescents, their families and the professionals.

6. Conclusion

There was no significant difference in PAL-value between boys and girls and there was a slight difference in TEE-value estimated with the activity diary and measured with the accelerometer. This study gives a well-researched background on which measurement methods are used in CF research today and also which limitations and problems arise when conducting such a research. Finally, authors see a need for further research in developing a platform for recommendations regarding physical activity as a part of the CF-care.
7. References


Appendix 1

Diagram 1 and 2. Whiskers plot regarding PAL-values taken from the activity diary (AD) respective the accelerometer (AC).
Appendix 2
This is the activity diary the participants where given, but translated into Swedish.

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Appendix 3

Information sheet given to the participants*

Information inför ert besök på energimetaboliska laboratoriet.
Ingång 95 nbv, Pediatrisk forskningsavd, Klinisk nutrition och metabolism, UAS.

För att undersöka kroppssammansättning och energiomsättning i vila och vid fysisk aktivitet används olika undersökningsmetoder. Undersökningarna görs för att få ett underlag till dina rekommendationer. Vi vill med denna information berätta om hur undersökningarna kommer att gå till. Vid undersökningen uppskattar vi om du kommer klädd så att det blir lätt att genomföra undersökningarna (dvs undvik strumpbyxor, tajta tröjor och klänning men ta gärna med gymnastikskor).

De undersökningar som är aktuella för dig är markerat med ett X i rutorna nedan:

1. **Mätning av energiomsättning i vila (grundförbrukning).**


2. **Undersökning av kroppssammansättning** görs med enkla metoder som mätning av vikt, längd och kroppsomfång (måttbandsmätning). Även andra metoder används:

   a) Bioimpedansmätning. **Hur?** Du får ligga på en bräde och ha elektroder på fingrarna eller handryggen och på fotterna. En mycket svag ström (som inte känns) passerar genom kroppen och det tar ca en minut. **Varför?** När det elektriska motståndet i vävnaderna mäts kan man ta reda på vilken mängd vatten som finns i kroppen.


Sammantaget ger undersökningarna ett mått på mängden underhudsfett.

3. **Undersökning av fysisk aktivitet** (ta med gymnastikskor).

a) **Gångprov i självald gånghastighet.**


b) **Konditions eller belastningstest.**

Konditionstest = 6‐minuters gångtest ☐ Belastningstest = Cykelergometer-test ☐


c) **Aktivitetsregistrering.**

**Hur?** Du får fylla i en dagbok under fyra dagar där du skattar de dagliga aktiviteternas intensitet på en niogradig skala. Aktivitetsregistreringen sker i samband med att du har på dig rörelsemätare (se nedan). **Varför?** De fysiska aktiviteten läggs samman med din grundförbrukning (se indirect kalorimetri) och tillsammans får vi en uppfattning om din totala energiomsättning. Aktivitetsdagboken ger information om vad och när du utför aktiviteterna.

d) **Rörelsemätare (accelerometer).**

**Hur?** Små mätare som bärs på hand- och/eller fotled och registrerar rörelse, acceleration och intensitet. **Varför?** Mätorna lagrar information om din fysiska aktivitet som spelas av efter avslutad registrering och visas i form av staplediagram. Du får ha på dig mätarna samtidigt som du fyller i din aktivitetsdagbok.

4. **Energiintag mha. kostdagbok.**

**Hur?** Om en analys av den totala energiomsättningen och din energibalans ska ske fylls även en kostdagbok i parallell med aktivitetsregistrering. Kostdagboken ger information om hur mycket energi som du får i dig, när du äter och hur ofta. **Varför?** Energiintaget ställs mot din fysiska aktivitet och därmed kan man räkna ut om du befinner dig i energibalans, dvs om du får i dig lika mycket energi som din kropp gör av med.

5. **Glukosbelastning**

**Hur?** En venkateter (plastslang) sätts in i ett ytliga blodkärl i ditt armveck. Därefter kommer blod att tappas från katetem före du dricker en sockerlösning och även 30, 60, 90, 120 minuter efter att du druckit den. Samtidigt görs en mätning av din energiomsättning då du får ligga med en plastkupa över huvudet (se även punkt 1).

**Varför?** För att undersöka din kropps förmåga att normalisera blodsockernivån och omsätta energi, kolhydrater och fett.

Om ni har frågor inför er undersökning kan ni kontakta:
Roger Olsson, näringsfysiolog, Linda Bratteby Tollerz, leg sjukgymnast eller Mia Berglund, forskningsassistent vid det energimetaboliska laboratori, UAS.
Telefon: 018-611 92 19.

*Compressed text taken from an information brochure, no changes have been made.*
Appendix 4

An illustration of affected organs in CF

Source: http://medical-dictionary.thefreedictionary.com/cystic+fibrosis