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Shwachman–Kulczycki score and resting energy expenditure in cystic fibrosis

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Abstract

Background: Disease severity assessed by clinical scores in cystic fibrosis (CF) has been a topic of investigation for many years, although a correlation of clinical scores with resting energy expenditure (REE) has not been described yet. We aimed to assess disease severity as evaluated by the Shwachman–Kulczycki (SK) score and to correlate these findings with REE and forced expiratory volume in 1 s (FEV_1). *Methods*: Twenty-eight patients performed respiratory function testing (FEV_1), and assessment of REE with open circuit indirect calorimetry. The SK score was evaluated according to general activity, physical examination, nutrition and conventional chest X-ray findings. *Results*: Mean SK score was 75.3 ± 15.7. Mean REE was 109.1% of predicted vs. 96.5% predicted in 16 healthy subjects ($P=0.002$). There was a significant correlation between the SK score and REE ($P=0.001$), the SK score and FEV_1 ($P<0.001$) and REE and FEV_1 ($P=0.034$). *Conclusions*: The correlations between the SK score, REE and FEV_1 demonstrate a close connection between disease severity, caloric requirement and lung damage. They confirm the clinical value of the SK score, which is easy to assess in a clinical setting.

Keywords: Nutrition; Calorimetry; Lung disease; Disease progression; Respiratory function tests

1. Introduction

Maldigestion due to exocrine pancreatic insufficiency is a major clinical manifestation in cystic fibrosis (CF). It leads to massive fat and protein losses, and deterioration of the nutritional status ensues. Therefore, optimal nutrition is a major cornerstone in the management of CF, and energy intake of at least 120% of the estimated average requirement (EAR) is commonly advised in patients with CF [1]. The actual caloric need, however, is most exactly assessed by measuring total energy expenditure (TEE), though indirect calorimetry is a more convenient method. TEE can be estimated from resting energy expenditure (REE), which has been shown to account for 60–75% of TEE [2].

The raised energy requirements in patients with CF are caused by repetitive lung infections, lung inflammation, increased respiratory work and more uncertain, the underlying genetic abnormality [3]. Good nutritional status is important in patients with CF, and long-term follow-up of CF patients has shown significantly better survival in patients who achieve normal nutritional status [4]. Disease severity assessed with clinical scores in CF has been topic of decade's of investigation [5], though a correlation of clinical scores with REE has not been described yet. The aim of the present study was to evaluate the validity of the Shwachman–Kulczycki (SK) score by correlating the score to REE and forced expiratory volume in 1 s (FEV_1). We also wanted to see whether these correlations were different in our paediatric compared to the adult patients with CF.

2. Materials and methods

Haukeland University Hospital serves as a regional centre for patients with CF in western Norway. We prospectively examined 13 female and 15 male patients of age more than five years from our cohort of patients with CF. The median age was 12.5 years (mean 14.7

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years, S.D. 8.4). The CF diagnosis was made by demonstration of repeated elevated sweat chloride concentrations (mean 100.1 mmol/l, S.D. 25.1) and typical clinical manifestations in all patients. The mean forced vital capacity was 95.3% of predicted (S.D. 22.7); the mean forced expiratory volume in 1 s was 80.8% of predicted (S.D. 23.2). Nine patients were found to be homozygous for the dF508 mutation (32%); the remaining 19 patients either had two other mutations (six patients), one detectable mutation (five patients) or no detectable mutation (eight patients).

Our patients attend the outpatient department every 2nd- 3rd-month, or more often when needed. Eight patients (29%) have chronic lung infection with *Pseudomonas aeruginosa.* Twenty-one out of 28 patients (75%) have been taking pancreatic enzyme supplementation (lipase 5000–10 000 IE/kg body weight).

All patients had received conventional therapy for CF for at least 6 months and they were in clinically stable condition. Informed consent was obtained from patients and/or their families. The regional ethical committee approved our study protocol.

Clinical status was evaluated by the SK score [6]. A maximum of 25 points and a minimum of one point are awarded on a five-point scale to each of the following categories: general activity, physical examination, nutritional status and conventional chest X-ray findings. This results in a global score from 4–100.

We performed standard computerised open-circuit indirect calorimetry (Sensor Medics Vmax, California, USA) after 12 h fasting, with at least four h restriction of using β-2 inhalative agents. The patients were placed under the canopy hood in a relaxed supine position. Respiratory gas exchange was monitored for 10 min to allow acclimatisation, followed by a subsequent 20–30 min measurement period. REE was calculated using the abbreviated Weir formula [7] on a steady state period. The steady state period was defined by a period of at least five consecutive minutes where oxygen uptake and ventilation varied by less than 10%, and respiratory quotient varied by less than 5%.

Standard spirometry was assessed by Sensor Medics Vmax (California, USA) equipment. We also examined 16 healthy subjects (seven females, nine males, median age 12 years, range 8–33 years) with indirect calorimetry in order to verify our predicted values of REE.

Data analysis was performed using a commercially available software package (SPSS). Descriptive statistics included mean, minimum and maximum values, standard deviations and median. Differences in REE (percentage of predicted) between healthy subjects and CF patients were tested using a Student's t test and a Wilcoxon–Mann–Whitney test. The relations between SK score, FEV_1 (percentage of predicted) and REE (percentage of predicted) were assessed by the Pearson

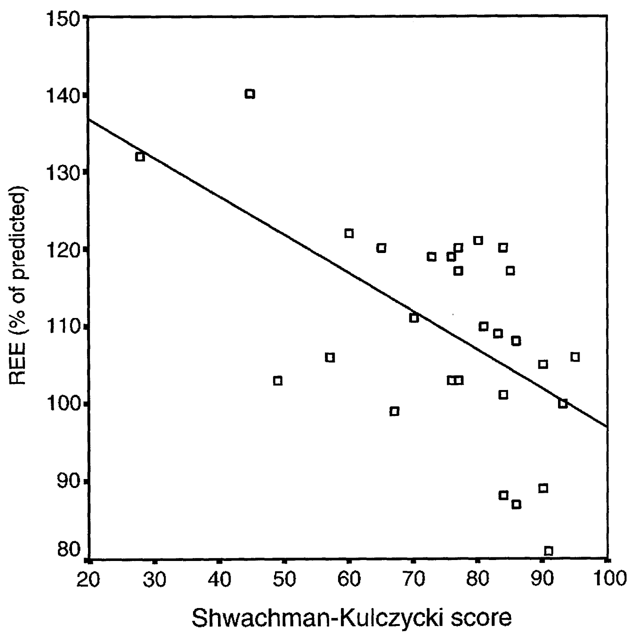

Fig. 1. Correlation between the Shwachman–Kulczycki score and resting energy expenditure (REE, percentage of predicted) in 28 patients with CF; the graph reflects a regression line. $R = -0.576$, $P = 0.001$.

correlation coefficient. All reported P values are two-tailed and $P < 0.05$ was considered significant.

3. Results

The overall mean SK score for our CF population was 75.3±15.7 (median 78.5), range 28–95. Mean value of resting energy expenditure was 109.1±13.6% of predicted in our CF population. In healthy subjects a mean value of 96.5±8.7% predicted was found, which was significantly lower ($P = 0.002$).

There was a significant correlation between the SK score and REE (percentage of predicted, $r = -0.576$, $P = 0.001$, Fig. 1). High resting energy expenditure was correlated with a low SK score.

Furthermore, there was a significant correlation between REE (percentage of predicted) and FEV_1 (percentage of predicted, $r = -0.403$, $P = 0.034$, Fig. 2). High resting energy expenditure was correlated with low FEV_1.

Additionally, there was a highly significant correlation between the SK score and FEV_1 (percentage of predicted, $r = 0.796$, $P < 0.001$, Fig. 3). Low SK score was correlated with low FEV_1.

In our younger patients ($N = 18$, age <16 years) the mean SK score was 80.1 (S.D. 11.2) compared to 66.8 (S.D. 19.5) in patients above 16 years of age ($P = 0.03$). There was no significant correlation between REE (percentage of predicted) and the SK score in our young patients, whereas, a significant correlation of FEV_1

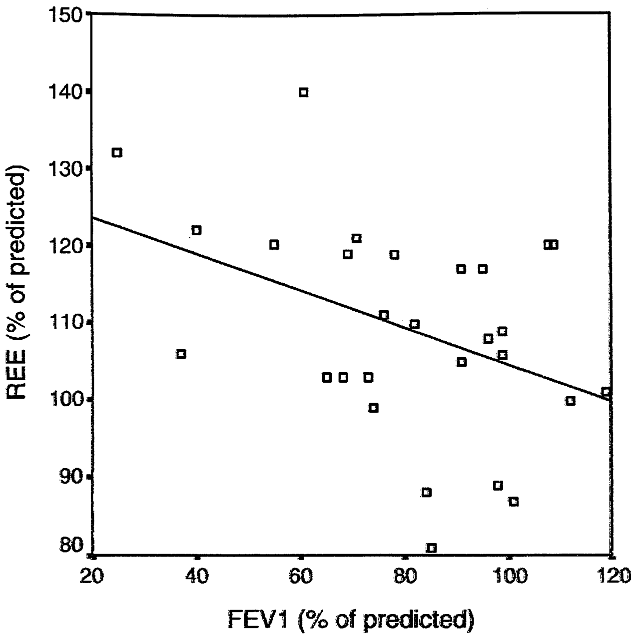

Fig. 2. Correlation between resting energy expenditure (REE, percentage of predicted) and forced expiratory volume in 1 s (FEV_1, percentage of predicted) in 28 patients with CF; the graph reflects a regression line. $R=-0.403$, $P=0.034$.

(percentage of predicted) and the SK score ($r=0.656$, $P=0.003$) was demonstrated in this group.

4. Discussion

The Shwachman–Kulczycki score has been used for almost half a century to assess disease severity in patients with CF, and it is still the most common clinical CF score [8,9]. Like other clinical CF scores [10,11] the SK score aims to provide easy accessible evaluation of a patient's overall condition according to the main categories general activity, physical examination, nutritional status and conventional chest X-ray findings. Although none of these items allow precise evaluation alone, the context of all items together gives information about the most life-limiting factors in CF, the degree of pulmonary destruction and malnutrition.

In our patients, we found a statistical highly significant correlation between the SK score and both REE and lung function as measured by FEV_1. Moreover, REE correlated significantly with FEV_1, which is in accordance with previously reported findings by Fried and co-workers [12]. In their study, they found a strong correlation between declining pulmonary function and increased resting energy expenditure. Once FEV_1 falls below 75% of predicted values, the REE raises in a curvilinear fashion. Buchdahl and colleagues demonstrated a significant correlation between increased REE and impaired pulmonary function as measured by the ratio of FEV_1 to FVC (forced vital capacity) [3]. In well-nourished CF males with well-preserved lung function, there is only little, if any increase in REE [12], although Bowler et al. found increased REE in CF patients with mild lung involvement as well [13]. Thus, disease severity seems to have an impact on resting energy expenditure. We have previously presented data on high-resolution computer tomography (HRCT) findings in our CF patients, and we found a significant correlation between a commonly used HRCT score and REE [14]. We could also confirm a significant correlation between HRCT and the SK score both in our paediatric and the adult patients (data not presented here) as reported by Demirkazik and colleagues [15]. Other investigators, however, could not find a correlation between REE and pulmonary function as measured by FEV_1 [16,17].

In the present study, the mean SK score in our CF population was 75, indicating mild to moderate disease. Significantly lower values were found in patients older than 16 years, reflecting more advanced disease in the adult CF population. The correlation between SK score and resting energy expenditure was, however, not significant for patients younger than 16 years. However, the correlation between SK score and FEV_1 was highly significant in this group. Taken into account our previous findings that HRCT score and SK were strongly correlated regardless of age, this lack of statistically significance among our younger patients may indicate a minor impact of mild disease on resting energy expenditure in

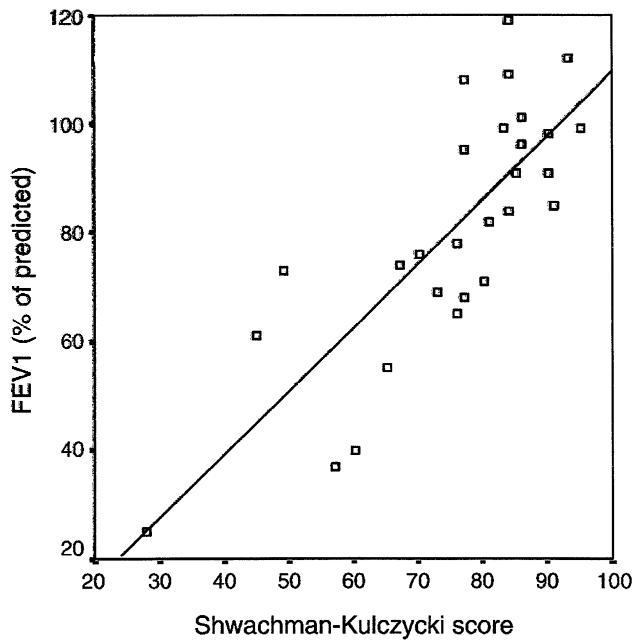

Fig. 3. Correlation between the Shwachman–Kulczycki score and forced expiratory volume in 1 s (FEV_1, percentage of predicted) in 28 patients with CF; the graph reflects a regression line. $R=0.796$, $P<0.001$.

CF. However, these findings must be interpreted with caution due to small sample sizes.

Increased REE in patients with CF is well known [4,18] and probably reflects repetitive lung infections, lung inflammation with elevated levels of cytokines and increased respiratory effort. The genetic defect itself may have a minor effect on REE, although recent studies have not revealed a primary defect of energy metabolism in subjects with CF [19,20]. The degree of parenchymal lung damage seems to play a major role in the interaction between REE and the SK score, which thereby confirms the findings of Fried and co-workers [12].

There seems to be a close correlation between disease severity evaluated by means of the SK score and resting energy expenditure. Similar correlations have been reported between the SK score and high-resolution computer tomography findings [15]. Our results emphasise the clinical value of the SK score, which in contrast to the other methods is easy to assess. The use of the SK score in evaluation of disease severity and progression in CF is still valid, especially in older patients.

Acknowledgments

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