

# Dietary intake and nutritional status in a Scandinavian adult cystic fibrosis-population compared with recommendations

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## Abstract

**Background:** Malnutrition is a well-known complication in cystic fibrosis (CF). There is good evidence that maintaining a normal body-weight correlates well with improved survival in CF. Energy intake in excess of 120% of the estimated average requirement (EAR) has been advised since 1980s.

**Objectives:** To investigate the nutritional intake and status in the adult Scandinavian CF-population.

**Subjects/Methods:** A cross-sectional multi-centre study was used to investigate the nutritional status of 456 adult CF-patients (2003–2006). Height and weight were measured and body mass index (BMI) and z-scores were calculated. Pulmonary function was examined by dynamic spirometry. A 7-day pre-coded food record (FR) obtained energy and nutrient intake data in 180 patients.

**Results:** The mean energy intake was 114 (SD 30.0)% of EAR and thus significantly lower than the target of 120% EAR ( $p < 0.001$ ) for patients with pancreatic insufficiency (PI) ( $n = 136$ ). Mean BMI was 22.0 (SD 2.9), the prevalence of BMI  $< 18$  was 13% and the prevalence of BMI  $\geq 25$  was 15% ( $n = 136$ ). Mean BMI was 20.8 (SD 2.4) in PI-patients with FEV<sub>1</sub>  $< 70\%$  and 23.2% (SD 3.0), in PI-patients with FEV<sub>1</sub>  $\geq 70\%$ , mean difference 2.4, (95% CI: 1.5, 3.3) ( $p < 0.001$ ), but there was no difference in energy intake. BMI  $\geq 18.5$  and a reported energy intake  $< 120\%$  were revealed in 54% of the PI-patients.

**Conclusions:** The energy intake did not reach the recommended 120% EAR, but the prevalence of underweight was lower than reported in other studies. The recommendation may exceed the requirement for a number of CF-patients. The nutritional status must still be closely monitored and nutritional advice and intervention should be individualised and adjusted to actual needs.

Keywords: *cystic fibrosis; adult; body mass index; energy intake; nutritional requirement; forced expiratory volume*

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Cystic fibrosis (CF) is an autosomal recessive genetic life-shortening disorder. Survival has

improved over time. A UK model predicts an estimated median survival of 50 years for a child born with CF in 2000 or later (1). In 2002, predicted survival at age 25 years was 95% for Swedish patients born in 1991 or later (2). The mean incidence of CF in Sweden is calculated to

For the Scandinavian Cystic Fibrosis Study Consortium (SCFSC), members are listed at the end of the paper.

be one in 5,600 live births and in Denmark one in 4,700 (3). The incidence in Norway is not known.

CF is characterised by multi-organ involvement primarily affecting the lungs and the digestive tract (4). Approximately 85% of patients with CF have pancreatic insufficiency (PI) leading to maldigestion and malabsorption (4, 5). Fat absorption between 85 and 95% of intake should be achievable with present enzyme preparations, but a substantial number of patients may not achieve this level of absorption (5).

Respiratory exacerbations and declining pulmonary function are major factors associated with increased resting energy expenditure (REE) in CF-patients (4, 6–9). The relationship between REE and pulmonary function suggests that an increase in REE occurs when forced expiratory volume in one second ( $FEV_1$ ) falls below 70% predicted value (6). The American CF Adult Care Consensus defines  $FEV_1 \geq 70\%$  of predicted value as normal or mild lung dysfunction (10). Additional findings support the notion that genotype and PI may be determinants of REE (7, 11, 12).

CF-related diabetes (CFRD) has been shown to worsen pulmonary function and nutritional status (13), although new data showed no difference in BMI between patients with and without CFRD (14).

The loss of nutrients and energy, and the increased REE result in higher nutritional needs. Accordingly, malnutrition is a well-known complication in subjects with CF leading to low weight, stunting, osteoporosis, essential fatty acid deficiency and low serum levels of the fat-soluble vitamins (4, 15, 16). The importance of nutritional status for survival of CF-patients is well documented and nutritional support is one of the cornerstones in the treatment of CF (7, 15, 17–19).

In a European consensus report, malnutrition is defined as body mass index (BMI) below 18.5 in adults >18 years of age (4). The American CF Adult Care Consensus defines malnutrition as BMI <19 (10). The Cystic Fibrosis Foundation recommends maintaining BMI  $\geq 22$  in females and BMI  $\geq 23$  in males aged 20 years and older, respectively (20).

Although malnutrition has been the main cause for nutritional concern in CF, overweight also exists. The prevalence of overweight and obesity (BMI >25) in adults with CF homozygous for the deltaF508 mutation was 10% based on data from the UK CF database in 2002 (21). As survival in CF increases, the potential risk of high BMI in adults may need concern.

Energy intake in excess of 120% estimated average requirement, (EAR) for energy has been advised for CF-patients (4, 7, 22). To achieve this level of energy intake, a high-fat diet is commonly advised (4, 15), usually more than 35% of energy (E%) from fat (7, 10).

Most reports of dietary intake in CF-patients focus on children, although there are a few reports focusing on energy and nutrient intakes in adults (23–26).

### Aim of the study

We investigated the energy intake and nutritional status, indicated by BMI, in the adult Scandinavian CF-population, with emphasis on PI patients.

We also investigated if PI-patients reached the recommended energy intake for CF of 120%–150% EAR of Nordic Nutrition Recommendations (NNR) (27). Furthermore, we wanted to find out if there were differences in the energy intake between PI- and PS-patients, between patients with normal and decreased lung function and between patients with and without CFRD.

### Materials and methods

#### Study population

This was a cross-sectional, multi-centre study, initiated and designed by the Scandinavian Cystic Fibrosis Study Consortium (SCFSC), investigating the nutritional status of the CF patients in Denmark, Norway and Sweden.

The Regional Ethics Committees in each country approved the study, and informed written consent was obtained from all patients.

Patients were included consecutively from September 2003 to May 2006, when in clinically stable condition. Inclusion criteria were adults ( $\geq 18$  years of age) with confirmed CF diagnosis based on clinical features, repeatedly ( $\geq$  two) positive sweat tests (chloride, Cl  $> 60$  mmole/l) and/or the presence of a known disease-causing mutation on each CFTR gene. Pregnancy was the only exclusion criterion.

Patients using pancreatic enzymes were defined as pancreatic insufficient. Patients using insulin or oral anti-diabetics were defined as having CFRD (14). Pulmonary function was examined at inclusion by dynamic spirometry at each centre. From measured forced vital capacity and expiratory volume in 1 second ( $FEV_1$ ) in litres, percentage of predicted values was calculated using Solymar and Quanjer reference equations for patients <19 and  $\geq 19$  years, respectively (28, 29).  $FEV_1 > 70\%$  predicted was defined as normal or mild lung dysfunction (10). Weight was measured in the morning wearing undergarments. Height was measured with no stockings or shoes and the means of three measurements were recorded. BMI was calculated. For calculation of BMI *z*-score, the reference values of Nysom were used (30).

Seven of the eight CF-centres in Denmark, Norway and Sweden participated with a total of 456 patients  $\geq 18$  years (Denmark 131 patients, Norway 83 patients, Sweden 242 patients). Food recording (FR) was done in

patients in all centres except the CF-centre in Gothenburg due to lack of resources. Demographic data exist for all the 456 patients and nutritional data exist for 180 patients (40%). The percentage of patients completing FR varied between the centres (Copenhagen 21%, Bergen 54%, Oslo 70%, Lund 59%, Stockholm 67% and Uppsala 14%).

Patients post-lung transplant ( $n=30$ ) were excluded from the calculations in this article.

Of the remaining 426 patients, 347 patients were pancreatic insufficient (136 patients with FR) and 79 patients were pancreatic sufficient (38 patients with food records, FR).

#### Assessment of dietary intake

A 7-day FR obtained energy and nutrient intake data. Nationally designed pre-coded forms were used, together with household measures and photographs to describe food portions sizes to assess the entire diet over 7 days. Seven days were chosen to eliminate bias related to different intake on weekdays and weekends. The Swedish and Danish pre-coded forms are validated (31, 32), whereas the validation of the Norwegian form has not yet been published. A CF dietitian instructed the patients how to complete the FR. The records were coded and analysed for energy and nutrients. In Sweden, this was done at the National Food Administration using the software MATs (Rudans Lättdata) with the database PC-Kost, in Denmark at Danish National Food Institute using the GIES software and in Norway at the Department of Nutrition, University of Oslo, using the KBS software. The calculation of energy was standardised using the factor 17 kJ for protein and carbohydrate (exclusive fibre) and 37 kJ for fat.

The reported energy and nutrient intake is the sum of food intake and oral- enteral energy supplements. Intake of vitamins, minerals and trace elements from supplementation was collected, and will be reported separately.

The energy intake as a percentage of EAR was calculated using reference values for weight and physical activity level (PAL: total energy expenditure divided by basal metabolism). PAL equal to 1.6, corresponding to sedentary work and limited physical activity in leisure time, was used (27).

#### Statistics

Continuous variables are described by means and standard deviations (SD) and categorical variables by counts (per cent). The one- and two-sample *t*-test and one-way ANOVA were used to compare continuous variables. The distributions of the continuous variables were determined to be sufficiently normal for the parametric tests to be appropriate (33). To compare categorical variables,  $2 \times 2$  tables were formed for each comparison. All the expected counts were above five and Pearson's chi-squared test was used in all cases. Statistical significance was set at  $p < 0.05$ . The analyses were performed using SPSS version 16.

## Results

#### Patients with pancreatic insufficiency (PI)

Data for 347 PI-patients are shown in Table 1. Patients completing FR, 136/347 (39%), were 2.8 years older and had a higher BMI than the patients not completing the FR. Both groups had normal BMI, and BMI *z*-score and lung function did not differ between the groups.

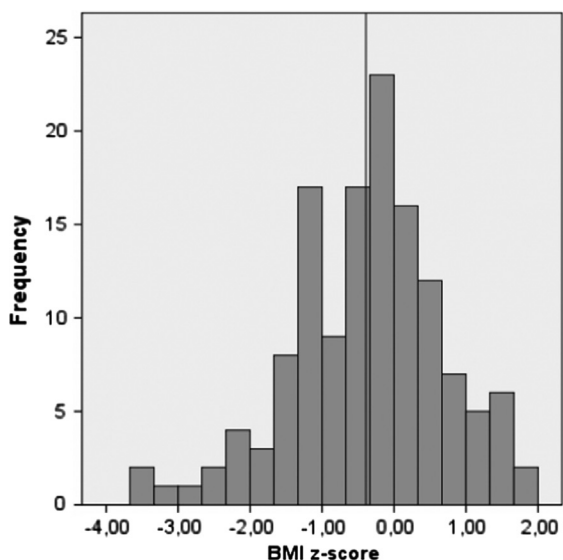
**Table 1.** Demographic and clinical data for 347 adult patients with cystic fibrosis and pancreatic insufficiency

	All patients $n=347$ Mean (SD)	FR $n=136$ Mean (SD)	No FR $n=211$ Mean (SD)	<i>p</i> -values
Age, years	29.6 (8.5)	31.3 (9.8) <sup>a</sup>	28.5 (7.3) <sup>a</sup>	0.005
FEV <sub>1</sub> , % predicted	68.0 (25.3)	69.4 (26.3)	67.2 (24.6)	0.43
FVC, % predicted	87.0 (21.9)	87.0 (22.4)	87.0 (21.6)	0.98
BMI, kg/m <sup>2</sup>	21.6 (2.9)	22.0 (2.9) <sup>b</sup>	21.4 (2.9) <sup>b</sup>	0.042
BMI <i>z</i> -score	-0.51 (1.1)	-0.39 (1.1)	-0.59 (1.2)	0.10
	<i>N</i> (%)	<i>N</i> (%)	<i>N</i> (%)	
Female	152 (44)	67 (49)	85 (40)	0.10
Diabetes mellitus (DM)	60 (17)	28 (21)	32 (15)	0.19
FEV <sub>1</sub> <70% of expected	178 (52)	67 (50)	111 (53)	0.54
BMI <19.0	62 (18)	22 (16)	40 (19)	0.51
BMI <18.5	44 (13)	17 (13)	27 (13)	0.94
BMI ≥25.0	39 (11)	20 (15)	19 (9)	0.10

Note: FR: food records.

<sup>a</sup>Age difference: 2.8, 95% CI (0.88, 4.7).

<sup>b</sup>BMI difference: 0.6, 95% CI (0.024, 1.3).



**Fig. 1.** Distribution of BMI z-score in PI-patients with food record. Mean value =  $-0.39$ .

Most PI-patients completing FR had normal BMI (18.5–24.9), 13% were underweight and 15% were overweight or obese.

BMI  $\geq 22$  was achieved in 21/67 (31%) females, and BMI  $\geq 23$  was achieved in 35/69 (50%) males. Fig. 1 shows the distribution of BMI z-score. The correlation between age and BMI z-score was small, but negative.  $R = -0.09$  ( $p = 0.08$ ).

The intake of energy, macro- and micronutrients for 136 PI-patients with FR is shown in Table 2. The mean intake for nearly all calculated nutrients reached the recommendations of NNR. The mean energy intake was less than the recommended energy intake of 120% EAR ( $p < 0.001$ ), and the target level was only achieved by 51/136 (38%) PI-patients. Fig. 2 shows the distribution of energy intake (% EAR).

No significant gender differences were observed for the variables energy intake (% EAR), FEV<sub>1</sub>%, FVC% or BMI z-score (data not shown). Mean BMI was 22.5

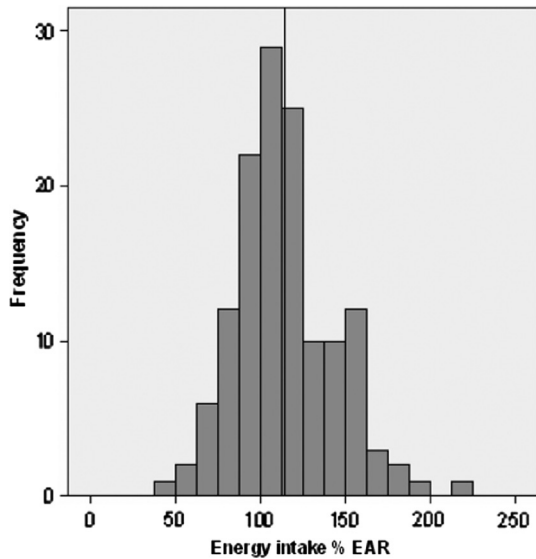
**Table 2.** Energy and nutrient intake from 7-day food records in 136 adult PI-patients

Energy and nutrients <sup>a</sup>	Intake $n = 136$ Mean (SD)	Recommended intake Energy and macronutrients	$p$ -values
Energy intake, % EAR	114 (30)	120–150 <sup>b</sup>	<0.001
Protein, E%	15.0 (2.3)	10–20 <sup>c</sup>	
Fat, E%	33.7 (6.3)	>35 <sup>b</sup>	0.015
–PUFA, E%	5.4 (2.2)	5–10 <sup>c</sup>	
Carbohydrates, E%	49.7 (6.7)	50–60 <sup>c</sup>	
–sugar, E%	13.3 (6.8)	<10 <sup>c</sup>	
Energy intake, kJ/d	12,159 (3,498)	–	
Protein, g/d	106 (31)	–	
Fat, g/d	112 (42)	–	
–PUFA, g/d	18.1 (10.4)	–	
Carbohydrates, g/d	354 (108)	–	
		Recommended intake Fibre and micronutrients <sup>c</sup>	
Fibre, g/d	19.6 (7.1)	25–35 g/d	
Thiamin, mg/d	1.8 (0.6)	F(females): 1.1, M(males): 1.4	
Riboflavin, mg/d	2.3 (0.9)	F: 1.3, M: 1.7	
Vitamin C, mg/d	121 (67)	F: 75, M: 75	
Calcium, mg/d	1,481 (627)	F: 800, M: 800	
Iron, mg/d	13.4 (5.1)	F: 15, M: 9	
Magnesium, mg/d	402 (130)	F: 280, M: 305	
Retinol equivalents, $\mu$ g/d	1,307 (732)	F: 700, M: 900	
Vitamin D3, $\mu$ g/d	5.8 (4.3)	F: 7.5, M: 7.5	
Tocopherol equivalents, mg /d	11.3 (5.9)	F: 8, M: 10	

<sup>a</sup>Intake of energy, macro- and micronutrients is calculated from food intake and oral and enteral energy supplements.

<sup>b</sup>Recommendations specific for CF (4, 17, 22).

<sup>c</sup>Nordic Nutrition Recommendations 2004 (27).



**Fig. 2.** Distribution of energy intake as a percentage of EAR in PI-patients. Mean value = 114%.

(SD 3.2) in males with FR and 21.5 (SD 2.5) in females with FR, a difference of 1.0, (95% CI 0.05, 2.0),  $p=0.004$ .

There was no correlation between energy intake (% EAR) and BMI or BMI  $z$ -score. The combination of a normal BMI ( $\geq 18.5$ ) and an energy intake less than 120% of EAR was found in 73/136 (54%) patients.

Mean E% from fat in PI-patients did not achieve the recommended level of 35 E% ( $p=0.015$ ) (Table 2). No differences in E% from fat between men and women were observed (data not shown).

#### *Patients with pancreatic sufficiency (PS)*

Demographic, clinical, and nutritional data were compared between PS- ( $n=38$ ) and PI-patients ( $n=136$ ) completing the FR. PS-patients were older, 37.3 years (SD 12.4) ( $p=0.008$ ), and had higher values of FEV<sub>1</sub>%, 79.6 (SD 20.0) ( $p=0.012$ ) and FVC%, 94.5 (SD 17.3),  $p=0.029$ . BMI and BMI  $z$ -score did not differ between the groups.

However, comparing all PS ( $n=78$ ) and all PI-patients ( $n=345$ ), mean BMI was significantly higher in PS, 23.2

(SD 4.4) versus 21.6 (SD 2.9) in PI-patients, a difference of 1.6, (95% CI 0.52, 2.6),  $p=0.004$ . Similarly, BMI  $z$ -score was  $-0.14$ (SD 1.3) in PS-versus  $-0.51$ (1.1) in PI-patients, a difference of 0.16, (95% CI 0.052, 0.68),  $p=0.023$ .

The mean energy intake (% EAR) was 104 (SD 23) in PS-patients, and this was significantly lower than 114 (SD 30) in PI-patients,  $p=0.04$ . The energy percentage from protein was 14.9 (SD 2.2), from fat 34.4 (SD 6.1), and from carbohydrates 47.9 (SD 6.2) and did not differ from PI-patients.

#### *PI-patients with reduced lung function*

We further analysed characteristics of PI-patients with FEV<sub>1</sub> < or  $\geq 70$ . BMI was significantly lower in patients with FEV<sub>1</sub> < 70% (Table 3). The difference in BMI = 2.4 (95% CI 1.5, 3.3) is equal to 7.0 kg based on a mean height of 171 cm in the PI-group ( $n=136$ ). Twelve patients (18%) were underweight (BMI < 18.5) compared to five patients (7%) with FEV<sub>1</sub>  $\geq 70\%$ ,  $p=0.06$ . There was a linear correlation between BMI and FEV<sub>1</sub>% predicted.  $R=0.367$ . BMI versus FEV<sub>1</sub>% predicted is shown in Fig. 3.

There was no significant difference between the groups regarding the percentage of energy from protein, fat or carbohydrate (data not shown).

#### *PI-patients with CFRD*

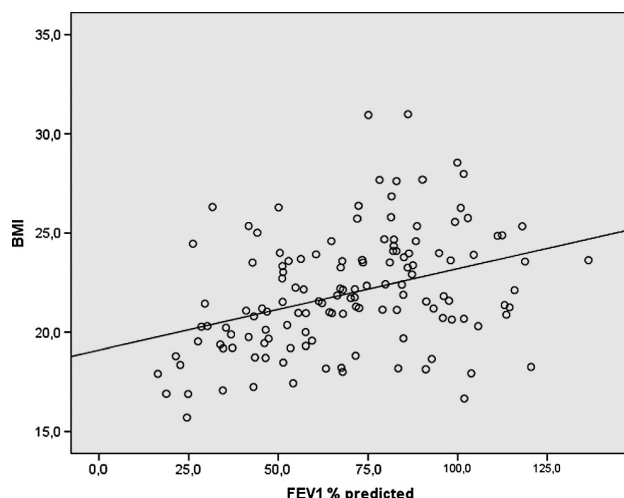
The prevalence of CFRD was 17% (60/347). When comparing all PI-patients with ( $n=60$ ) and without ( $n=487$ ) diabetes, we found no differences in BMI and BMI- $z$ -score.

Table 4 shows demographic and clinical data for patients with and without CFRD completing the FR. The diabetic PI-patients with FR had lower BMI, a more negative BMI  $z$ -score and a lower FEV<sub>1</sub>% predicted than the non-diabetic patients. We observed no significant difference in energy intake between the diabetic (107% EAR) and non-diabetic (116% EAR) PI patients. However, we found some differences in the intake of macronutrients, with higher E% from fat,  $p=0.032$ , and lower E% from carbohydrates,  $p=0.019$ , in diabetic patients (Table 4).

**Table 3.** Clinical and nutritional data in 135 PI patients with food records, grouped by FEV<sub>1</sub>% predicted

	FEV <sub>1</sub> < 70% $n=67$ Mean (SD)	FEV <sub>1</sub> $\geq 70\%$ $n=68$ Mean (SD)	$p$ -values
Age, years	32.6 (10.3)	30.0 (9.3)	0.13
FEV <sub>1</sub> , % predicted	47.4 (14.3)	91.1 (14.8)	–
FVC, % predicted	70.0 (16.3)	104 (13.0)	–
BMI, kg/m <sup>2</sup>	20.8 (2.4)	23.2 (3.0)	<0.001
BMI $z$ -score	$-0.88$ (1.0)	0.09 (0.9)	<0.001
Energy intake, % EAR	113 (30)	114 (30)	0.86





**Fig. 3.** Plot of BMI versus FEV<sub>1</sub>% predicted in PI-patients with food record. The equation for the regression line is  $BMI = 19.1 + 0.0415 \times FEV_1\% \text{ predicted}$ , indicating that an increase in FEV<sub>1</sub> of 1% is associated with an increase in BMI of 0.0415.

## Discussion

In this study, the dietary intake and nutritional status in the adult CF-population of Scandinavia, with emphasis on PI-patients, were investigated.

The prevalence of underweight was low in Scandinavia (13%). According to the 2005 CF Foundation Patient Registry Report, the prevalence of BMI < 18.5 was 22% in adult CF patients with PI in the USA (20). White et al. reported the prevalence of BMI < 19.0 to be 26% in 80 adult patients in Leeds who completed a 4-day unweighed dietary record in 1995–2000. Mean age was 23.8 (SD 6.4) years, mean FEV<sub>1</sub>% was 58.7 (SD 23.9) and mean BMI 20.8 (SD 2.6) (24). The higher prevalence of underweight in this study may be explained by the severity of lung disease. The prevalence of BMI ≥ 25 was 11% in our study when all PI-patients ( $n = 347$ ) were included. This

was comparable to the results from the UK CF database 2002. They reported the prevalence of BMI > 25 to be 10%, although mean FEV<sub>1</sub>% was lower than in the Scandinavian population (21). The quality of the dietary intake seemed satisfactory based on the mean intake of calculated nutrients (Table 2). Almost all nutrients were in accordance with recommendations in NNR (27).

Mean values for energy and fat intake are lower than reported in the studies of White et al., Richardson et al. and Oliveira et al. (23–25). Oliveira et al. reported the highest energy intake analysing a 7-day weighed FR from 37 (70% PI) Spanish patients > 16 years of age who had completed puberty. Mean age was 24.5 (SD 9) years, mean FEV<sub>1</sub>% 64 (SD 27) and mean BMI 21.8 (SD 4). BMI was < 18.5 in seven (19%) patients. The energy intake of the 37 patients was 132% of EAR, and E% from fat was 39% both for the patients and the 37 controls (25). Regardless of the high mean energy intake and including PS-patients in the group, their nutritional status was not better than in our study. The quite few patients in the Spanish study may be an explanation for this.

Oliveira et al. commented that a high energy, fat-rich diet is relatively easy to achieve in Spain because this is part of the ordinary dietary habits in the area. In our study, E% from fat was at the same level as reported from national dietary surveys in general population of Scandinavia (34).

It is not possible to tell from this Scandinavian cross-sectional study if the patients were in energy balance with the reported intake. Energy intakes as low as 50% EAR cannot represent the energy intake necessary for energy balance (Fig. 2). The patients who reported the lowest energy intakes may have underreported their real intake, or these intakes may be typical for a shorter period of time.

Collins et al. have reported a lower energy intake in females than in males (35). We found no significant

**Table 4.** Clinical and nutritional data in 136 PI patients with food records, grouped by diabetes

	PI and diabetes $n = 28$ Mean (SD)	PI without diabetes $n = 108$ Mean (SD)	$p$ -values
Age, years	31.5 (10)	31.2 (9.8)	0.89
FEV <sub>1</sub> , % predicted	60.8 (23.6)	71.6 (26.6)	0.04
FVC, % predicted	81.7 (24.5)	88.3 (21.7)	0.20
BMI, kg/m <sup>2</sup>	21.0 (2.7)	22.3 (3.0)	0.03
BMI-z-score	-0.77 (1.1)	-0.29 (1.0)	0.04
Energy intake, % EAR	107 (33)	116 (29)	0.21
Protein, E%	15.4 (2.4)	14.9 (2.3)	0.36
Fat, E%	36.0 (6.8)	33.0 (6.1)	0.03
Carbohydrates, E%	46.8 (7.1)	50.5 (6.4)	0.02

Note: BMI, body mass index; CF, cystic fibrosis; CFRD, CF related diabetes; EAR, estimated average requirement; E%, percent of energy intake; FEV<sub>1</sub>, forced expiratory volume in 1 second; FR, food record; FVC, forced vital capacity; NNR, Nordic Nutrition recommendations; PAL, physical activity level; PI, pancreatic insufficiency; PS, pancreatic sufficiency; PUFA, polyunsaturated fatty acids; REE, resting energy expenditure.

gender differences in energy intake (% EAR), BMI *z*-score or FEV<sub>1</sub>% predicted.

The difference in mean energy intake between PS- and PI-patients was probably reflecting a real difference in energy requirement due to normal pancreatic status and better lung function in PS-patients. It was surprising that BMI and BMI *z*-score did not differ between PS- and PI-patients completing FR. This may be by chance or may be due to PS-patients being concerned about their weight were more inclined to food recording.

A lower BMI and a more negative BMI-*z*-score in the group with reduced lung function revealed a larger gap between mean energy intake and energy requirement compared with the group with nearly normal lung function. This may be explained with an increase in REE, although an increase in REE does not always mean an increase in total energy expenditure (8). It is also possible that the patients with moderate or severe lung disease have been over reporting their intake. Under-reporting of energy intake is common with dietary self-reporting methods, but over-reporting of energy intake has been reported in under weight men (36). In a recent study by Oliveira et al., under-reporting in a 7-day FR was lower in CF patients than in controls (37). The authors discuss the possibility of over reporting due to awareness of nutritional status and energy intake.

The lower BMI and BMI *z*-score in the diabetic patients completing FR also revealed a larger gap between energy requirement and energy intake in the diabetics compared to non-diabetics. The dietary advices for CFRD have to take into account the other considerations of CF. Special attention should be given to avoid unnecessary restriction of carbohydrates and to the energy intake to prevent or treat weight loss (4, 10).

In the case of weight loss, the European consensus report recommends a full re-evaluation of all the possible causes that may affect nutritional status. If nutritional intervention is indicated, the first step should be maximising energy intake using energy-dense foods, then adding supplements and as a third step enteral nutrition may be indicated, usually over night (4). Our results indicate that there is still a potential for better nutritional follow-up of patients with CF in Scandinavia, especially when lung function decreases.

The low percentage of FR in two of the centres (Copenhagen and Uppsala) and no food recording in the centre in Gothenburg may have impaired the representativity of the FR-group for the whole cohort, although essential clinical data did not differ (Table 1). Dietary recording is burdensome, and volunteer bias may be a problem in all studies using dietary recording (32).

In conclusion, the mean energy intake did not achieve the recommendation in CF. The recommendation of energy intake in excess of 120% EAR is frequently

referred to in the CF-literature (4, 7, 22) but is based on quite crude estimates of energy loss and energy expenditure. The heterogeneity of CF-patients, including presence of PI, respiratory infection, reduced lung function, activity and nutritional status, means that universal recommendations may be difficult to give.

The prevalence of underweight was low in Scandinavia. Because CF-patients nowadays are doing far better than previously, the current recommendations for energy intake may exceed the requirement in a number of CF-patients.

However, BMI *z*-score is still negative compared to the general population data, and nutritional status should be closely monitored to prevent or treat unintentional weight loss.

With increased longevity, the potential risks of high BMI in CF need further investigation and concern. Nutritional advice and intervention should be individualised and adjusted to actual needs, and be appropriate to support long-term health.

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### Conflict of interest and funding

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### Scandinavian cystic fibrosis study consortium

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