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Dietary intakes in adult patients with cystic fibrosis–do they achieve guidelines?

H. White[a,*], A.M. Morton[a], D.G. Peckham[b], S.P. Conway[b]

[a]*Dietetic Department, Seacroft Hospital, York Road, Leeds, England, LS14 6UH, UK*
[b]*Regional Cystic Fibrosis Unit, Seacroft Hospital, York Road, Leeds, England LS14 6UH, UK*

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Abstract

Background: Most patients with cystic fibrosis (CF) require a higher energy and protein intake than their healthy peer group. There are few data on dietary intakes of adult patients. The aim of this study was to determine nutritional intakes in an adult population with CF. The impact of nutritional intervention and disease on macronutrient intake was examined. *Methods*: Retrospective cross-sectional analysis of 94 unweighed food diaries at annual review (1995–2000). Energy and protein intakes were compared to the estimated average requirement (EAR) for energy and reference nutrient intake (RNI) for protein. The effect of diet alone, oral supplements, enteral tube feeding, and cystic fibrosis related diabetes (CFRD), on macronutrient intake was examined and impact of pancreatic sufficency (PS) and lung transplantation. *Results*: Mean energy and protein intakes approached recommended CF guidelines, but in 72% of assessments these values were not achieved. Mean energy and protein intakes for patients on diet alone and protein intake for those with CFRD failed to meet recommendations. Oral supplementation and enteral tube feeding regimens increased energy and protein intake above recommended levels. No group achieved 40% total energy from fat. Patients receiving enteral tube feeds had the highest mean energy and protein intakes but lowest body mass index (BMI) and lung function. *Conclusion*: Adequate mean energy and protein intakes in adult patients with CF mask subgroups of patients who fail to meet recommendations ie. diet alone, diabetic. Oral supplementation and enteral tube feeding increase energy and protein intake but fail to achieve an adequate BMI level in subjects with a decreased clinical status. Individual nutritional assessment remains essential.

Keywords: Cystic fibrosis; Adult population; Nutritional intake

1. Introduction

The median age of survival with cystic fibrosis (CF) is now approximately 30 years [1]. Advances in dietary management with an emphasis on higher fat diets and the use of acid resistant pancreatic enzyme replacement therapy (PERT) has been important in contributing to better health, quality of life and longevity [2,3].

Most patients with CF have higher energy requirements than healthy individuals of the same age due to increased losses from fat malabsorption [4], increased demands imposed by infection and inflammation [5] and a reduction in dietary intake especially during episodes of infection. The adaptive response to malnutrition in CF lung disease may also be abnormal and a marked increase in muscle protein catabolism of patients with CF when compared to healthy controls has been shown [6]. The energy requirements of undernourished adolescents and young adults with cystic fibrosis are 25–80% higher than healthy individuals of the same age and size [7]. Nutritional guidelines, therefore recommend an intake for individuals with CF of 120–150% of the estimated average requirement (EAR) for energy and 200% of the reference nutrient intake (RNI) for protein [8,9]. The dietary aim is to encourage a diet high in fat (40% of total calories) and to maintain this even with disease progression. Studies in children indicate that the above can be difficult to achieve [10–12]. Richardson et al. (2000) [13] have reported the nutritional intake of an Australian adult population although

*Corresponding author..

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Table 1
Subject characteristics at assessment

	Assess (n)	Mean age (years)	Mean BMI (kg/m^2)	Mean FEV_1 (%)	Mean FVC (%)
Total*	80	23.8 ± 6.4	20.8 ± 2.6	58.7 ± 23.9	75.3 ± 24.2
Diet alone	27(34%)	22.0 ± 5.0	21.2 ± 2.6	65.5 ± 24.6	82.1 ± 24.8
Oral supps	14(17%)	21.0 ± 3.3	21.1 ± 2.3	66.2 ± 21.0	86.7 ± 21.6
Enteral feed	10(12%)	21.1 ± 3.3	18.5 ± 1.9	37.9 ± 13.4	52.4 ± 13.3
Diabetic	29(36%)	27.8 ± 7.5	21.1 ± 2.8	55.3 ± 23.5	70.7 ± 22.2

BMI, Body mass index; FEV1, Forced expiratory volume in 1 s; FVC, Forced vital capacity.
* PS, post-transplant excluded.

there are little other data on the nutritional intake of adults.

The aim of this study was to determine whether dietary intakes in an adult population attending a regional cystic fibrosis unit meet current recommended guidelines and whether defined aspects of nutritional intervention and disease influence energy and macronutrient intake.

2. Patients and methods

This retrospective cross-sectional study examined 94 dietary assessments (48 male) completed as part of the annual review at a time of clinical stability during the period 1995–2000. Height and weight were recorded on the day of assessment and BMI (weight/height m^2) calculated.

Pulmonary function was assessed by means of standard spirometry using a Vitalograph Compact II Spirometer (Vitalograph Ltd, UK). Individuals were required to perform 1–3 satisfactory efforts and the best measurements were recorded. 'Satisfactory' was defined as FEV_1 lying within 5% of last recorded respiratory function or repeated respiratory function on the same day. Forced expiratory volume in 1 s (FEV_1) and forced vital capacity (FVC) were compared with reference values and reported as the percentage of the predicted normal value.

At each assessment the presence of diabetes, PS and transplant status was recorded. Length of time post-transplant was noted. A diagnosis of diabetes mellitus was based on the need for insulin therapy or oral hypoglycaemic agents (pre 1995) or a positive glucose tolerance test with a clinical need for insulin therapy from 1995 onwards. Pancreatic insufficiency was defined as the presence of malabsorption requiring pancreatic enzyme replacement therapy (PERT). All patients routinely received advice on maximising dietary energy, protein and fat intake irrespective of BMI. Post-transplant patients received dietetic review a minimum of twice yearly, PS patients a minimum of 4 times/year and remaining patients a minimum of 6 times/year.

Each patient received instruction from one of two specialist dietitians on completing a 4 day un-weighed dietary food record inclusive of one weekend day. All food, drink, nutritional supplements enteral feeds and PERT were recorded. Date of initiation of enteral feeding was noted. Food records were analysed by the same specialist dietitians using Microdiet Computer software (University of Salford, UK, 1983–1995) to establish energy and macronutrient intakes. Energy and protein intakes were compared to the EAR for energy and RNI for protein [14]. Percentage energy contribution from protein, fat, carbohydrate and alcohol was calculated for each assessment.

Of the total number of assessments ($n=94$), those classified as pancreatic sufficient ($n=6$) and post-transplant ($n=8$) were excluded and analysed separately. The remaining assessments ($n=80$) were divided according to the consumption of diet alone, dietary supplements, enteral tube feeding, or the presence of CFRD, in order to determine any differences in energy and macronutrient intakes between these subgroups. The contribution of oral supplements and enteral feeds to total energy and protein intakes in these patient groups was calculated. Weight, BMI and dietary intake in different age categories was examined, excluding PS and post-transplant patients to avoid bias. PS and post-transplant groups were analysed separately for energy and macronutrient intake.

Results are expressed as mean values with the range or standard deviation. For comparison between groups two-tailed unpaired t tests were used. The Spearman rank linear correlation coefficient was calculated to describe the interrelations between two measurements. A P value of <0.05 was regarded as significant.

3. Results

3.1. Subject characteristics

Patient characteristics at assessment are detailed in Table 1. BMI was 21.2, 21.2, 18.5, 21.1 in patients on diet alone, oral supplements, enteral tube feeding and diabetic, respectively. Corresponding % predicted FEV_1 was 65.5, 66.2, 37.9 and 55.3%.

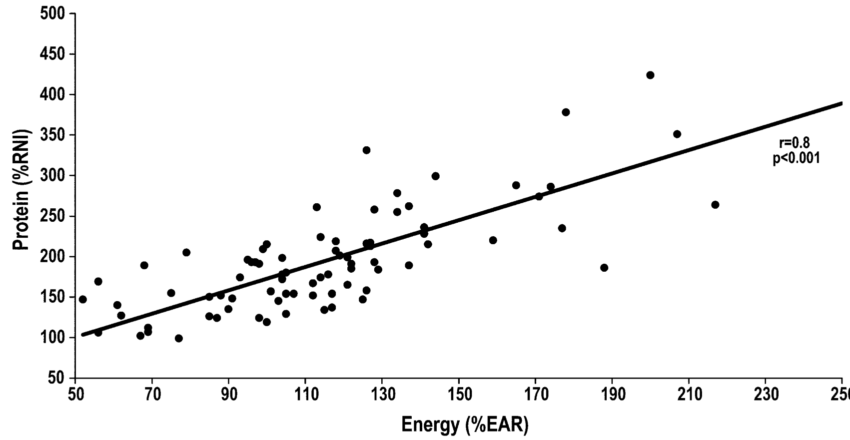

Fig. 1. Relationship between dietary energy and protein intake ($n=80$).

3.2. Energy and protein intakes

Mean energy intake (117% EAR) and mean protein intake (197% RNI) were close to recommended guidelines. There was a significant correlation between energy and protein intake ($r=0.8$, $P<0.01$) (Fig. 1). Although mean intakes were close to recommended levels, only 40% of all assessments achieved an energy intake of 120% EAR, 38% a protein intake of 200% RNI and only 28% achieved both objectives. In 21 (26%) assessments patients had a BMI <19 kg/m^2. Comparison of BMI and energy intake showed that 12(15%) had a BMI <19 kg/m^2 and energy intake $<120\%$. In 6(7.5%) assessments, patients had a BMI<19 kg/m^2 and energy intake $<100\%$ EAR.

Differences in mean energy and protein intakes for defined patient groups are shown in Figs. 2 and 3. These show a stepwise increase in energy and protein intake from those on diet alone to supplements to enteral tube feeding. With each nutritional intervention an increase in energy and protein intake was seen with respective figures of 100%, 126% ($P<0.01$) and 140% (NS) EAR for energy and 165, 221% ($P<0.01$) and 248% (NS) RNI for protein.

Diabetic assessments achieved a mean energy intake of 120% EAR energy and 197% RNI protein and almost achieved recommended intakes. Minimum current guidelines were achieved by the supplement group and enteral tube feed groups only.

3.3. Macronutrients

The recommended percentage dietary intake from fat is 40%. This was not achieved by either the group as a whole or by any patient subgroup (Table 2). The highest percentage fat intake was achieved by those on enteral feeds (38.7%) and the lowest by those who were diabetic or on diet alone (34.8%). Alcohol intake was low in all groups (Table 2) but decreased as patients progressed from diet alone to supplements and then to enteral feeding.

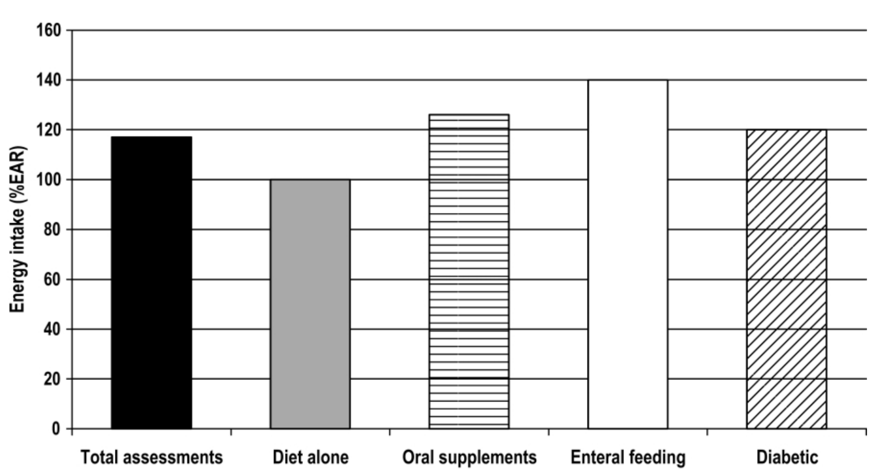

Fig. 2. Mean energy intake (% EAR).

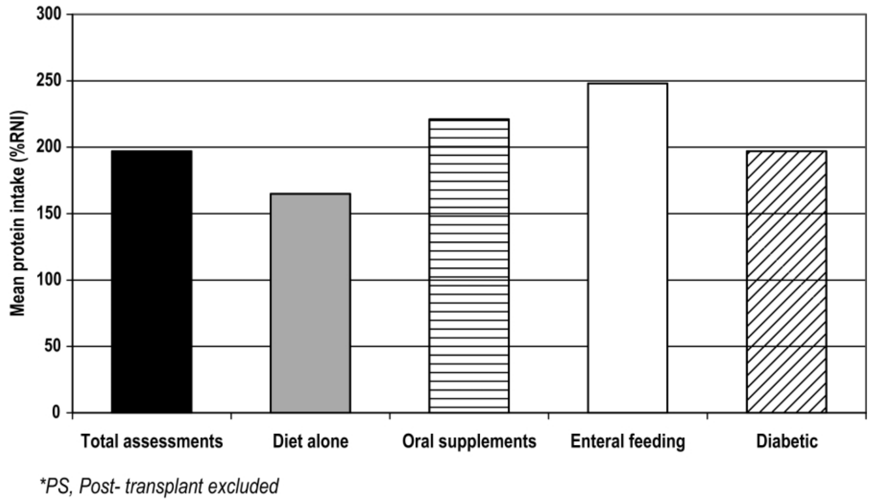

Fig. 3. Mean protein intake (% RNI).

Table 2
Macronutrient intake in adult patients with cystic fibrosis

Patient group	% Fat	% CHO	% Prot	% Alc
Total	35.7 ± 6.2	47.9 ± 6.2	15.2 ± 3.4	1.6 ± 3.3
Diet alone	34.8 ± 7.2	48.4 ± 7.7	14.9 ± 3.3	2.6 ± 4.6
Supplements	36.0 ± 4.9	47.2 ± 6.1	15.0 ± 3.5	1.8 ± 2.9
Enteral feed	38.7 ± 2.2	45.6 ± 3.6	15.5 ± 2.1	0.02 ± 0.06
Diabetic	34.8 ± 6.6	48.4 ± 5.6	15.6 ± 3.9	1.0 ± 2.1

Oral supplement use and enteral tube feeds: Oral supplements were used in 17.5% of assessments and enteral feeds in 12.5%. Contribution of oral supplements to overall energy intake was 17.3% and to protein intake 11.4%. Supplements were more commonly used in males than females (71% vs. 29%). It was noted that the younger age groups derived a higher percentage of their nutrient intake from oral supplements. Individuals taking oral supplements had a similar BMI and lung function to those on diet alone.

The median length of time that patients were enterally fed prior to assessment was 3.1 years. Enteral feeders relied heavily on tube feeds to maintain their high nutritional intakes of 140% EAR energy and 248% RNI protein. These provided 50.6% of total energy and 51.1% of protein intakes.

3.4. CFRD

Patients with CFRD were significantly older ($P < 0.001$) than non-diabetics and had reduced lung function although this did not reach significance. Diabetic patients met recommended energy guidelines but not protein guidelines (Fig. 3) or % energy from fat (Table 3).

3.5. Age

The data were examined to see if there was any effect of age on dietary intake (Table 3). Mean values of energy intake were higher in each age category until age 24. Those aged 20–24 also had the highest protein intake. BMI levels increased in the age categories 15–29, decreased at age 30–34 and increased again in the age categories 35–50. The two individuals in the age category 40–50 had the highest BMI but lowest energy intake of any group.

3.6. PS and post-transplant

Patients with PS had the greatest lung function and BMI of any group despite a low energy intake (104% EAR) and protein intake (175.8%) compared to guide-

Table 3
Change in weight, BMI and dietary intake with age

Age (years)	Assess (n)	Wt (kg)	BMI (wt/kg^2)	% EAR energy	% RNI protein	% E fat
15–19	29	53.6 ± 7.3	19.9 ± 2.0	111.6 ± 385	192.5 ± 78.1	36.1 ± 4.5
20–24	25	60.9 ± 11.5	21.3 ± 3.1	127.6 ± 40.4	212.7 ± 73.4	35.9 ± 6.1
25–29	14	60.4 ± 9.9	21.7 ± 2.1	111.4 ± 39.2	177.0 ± 51.3	33.3 ± 8.1
30–34	5	55.4 ± 6.4	19.9 ± 1.2	113.6 ± 35.7	211.4 ± 62.1	36.0 ± 11.5
35–39	5	60.8 ± 17.0	21.6 ± 4.5	116.2 ± 28.7	191.0 ± 52.0	34.0 ± 3.9
40–50	2	62.3 ± 17.9	22.5 ± 4.6	106.0 ± 30.0	181.5 ± 44.5	38.4 ± 3.4

Table 4
Demographic and nutritional data for pancreatic sufficient and post-transplant patients with cystic fibrosis.

	PS ($n=6$)	Post-transplant ($n=8$)
Mean age (years)	28.0±3.9	29.9±7.8
Mean BMI (kg/m^2)	25.1±2.8	21.0±4.2
Mean FEV_1 (%)	85.7±14.4	68.1±32.3
Mean FVC (%)	99±13.2	82.4±26.1
% EAR energy	104.2±31	94.8±17.9
% RNI protein	175.8±26.6	157.1±29.5
% energy fat	36.3±4.0	38.0±4.1

lines (Table 4). Post-transplant patients were the oldest group of patients within the study and had an adequate BMI despite the lowest mean energy intake (94.8% EAR). Mean length of time post-transplant was 4.7 years. There was a strong correlation between BMI and FEV_1 ($r=0.64$, $P<0.001$) in this group.

4. Discussion

Several studies have examined nutritional intake in younger or mixed age populations of people with CF [10,12,15,16] but there are little published data on the nutritional intakes of adults over 16 years, or whether nutritional guidelines are met or are influenced by changes in nutritional intervention and clinical aspects of the disease.

Mean energy and protein intakes achieved were close to recommended guidelines but less than the reported energy intake in an Australian population of adult patients with CF [13]. The mean of the group hides a large proportion who lie below recommended levels, i.e. 60% failed to achieve recommended energy intakes, 62% failed to achieve recommended protein intake and 72% failed to achieve both. This emphasises the need for individual dietetic assessment and care for all patients. A high fat diet (>40% total energy from fat) is an essential component of a high-energy intake. Groups with the lowest fat intake were those on diet alone who failed to meet energy and protein guidelines and diabetic patients who failed to meet recommended guidelines for protein. However, those consuming diet alone also demonstrated the highest mean BMI, FEV_1 and FVC% predicted of any patient (PS and post-transplant excluded)). Adequate nutritional status (BMI 21.2 kg/m^2) despite lower than recommended intakes indicates that current guidelines are inappropriately high for this comparatively well subgroup of the clinic population.

CFRD is associated with clinical deterioration [17–19] and may precede overt diabetes by up to 4 years. Emphasis on a high fat diet, with no restriction of refined carbohydrate, is thought to be important in achieving the higher energy intakes required in this more compromised group. This conflicts with dietary information given to the non-CF diabetic population where a diet low in refined carbohydrate, low in fat and high in fibre is advised. Patients with CFRD achieved recommended guidelines for energy but not % energy from fat. The higher % energy contribution from carbohydrate of the CFRD diet (48.4%) was closer to guidelines for the general diabetic population [20] and those on diet alone. Further studies are required to establish whether recommended changes in dietary intake occur at diagnosis or over time.

Work has shown that dietary intake may become compromised in CFRD because of problems concerning food and eating behaviour [21]. A consistency of approach and nutritional targeting is, therefore needed in this more vulnerable group of patients.

The use of oral supplements is the first line treatment when dietary counselling alone fails. In adolescents their use is associated with a significant increase in energy intake, but without any corresponding improvement in nutritional indices [16]. Approximately one-fifth of the assessments in this study included oral supplements that resulted in significant increases in energy and protein intake. Similar improvements in energy intake have previously been reported [16]. Oral supplements provided 17.3% of the total energy and 11.4% of the total protein intake. The greatest use of oral supplements was in the younger age groups 15–19 years (31%) and 20–24 years (40%). The gradual reduction in use thereafter may be due to a combination of taste fatigue, progression to enteral feeding and survival of milder genotypes requiring less intensive nutritional therapy.

Despite the highest mean energy and protein intakes and the highest percentage energy intake from fat, enteral tube feeders had the poorest lung function and BMI of any group within the study. Bell et al. (1998) [22] reported a similar trend in an adult Welsh population. The enteral tube feeds provided half of the energy and protein intake for these patients, although the choice of formulae contributed to the higher energy and protein intakes observed (patients routinely received a 2 kcal/ml high fat feed). There is a need for longitudinal data to show the effects of enteral tube feeding on nutritional status and lung function.

As severity of lung disease and nutritional status are related, nutritional status might be expected to decline with age. Nir et al. (1996) [3] examined long term survival and nutritional data in a Danish population and showed that weight and BMI progressively decreased in patients >15 years. None of their patients were enterally tube fed. In this study, mean weight was initially seen to increase as young, sicker individuals died and then decreased around the median age of survival. A further increase in weight and BMI was then observed in the older group of patients (aged 35 and over). This suggests that age and preservation of nutritional status are possibly related to a milder disease phenotype. Inclusion of such data in future studies may provide valuable additional information on dietary requirements.

Further limitations concern the collection of data in the study. Food diaries were obtained from patients attending for annual assessment and can be time consuming to record. The patient's willingness to complete and return the food diary could obviously introduce a bias in favour of more compliant or motivated individuals. Nonetheless the completed assessments were proportionally representative of the clinic population in terms of the percentage of patients consuming diet alone, oral supplements and enteral feed [23].

The unique characteristics of PS and post-transplant patients were evident in their nutritional status and dietary intake. Those with PS had a mean BMI lying above the desired BMI range of 19–25 for members of the general UK population. Lack of malabsorption and comparitively good lung function will potentially lower requirements in this patient group. The low energy intake (104% EAR) in PS patients indicates that their energy requirements are closer to that of the non-CF population. The requirements of CF individuals post-transplant are also unclear. Madill et al. (1993) [24] were able to improve nutritional status post-transplant with significantly decreased caloric intake. Our own data did not allow us to observe changes over time but demonstrates that an adequate BMI can be achieved post-transplant with a relatively low energy intake. Further work is needed to establish energy requirements and longitudinal changes in these groups.

5. Conclusion

In conclusion, we have shown that mean energy and protein intakes in an adult population are close to recommended guidelines, although fat recommendations are not met. Adequate mean energy and protein intakes mask the fact that $<40\%$ of patients achieve the recommended intake for energy or protein and only 28% achieve both. The mean hides an important undernourished group of patients (15%) who had a BMI <19 kg/m^2 and energy intake of $<120\%$ EAR.

Particular subgroups do not achieve recommended levels, i.e. those on diet alone who may have lower requirements and those with CFRD who appear to need greater dietary intervention and monitoring, although the latter group only just fell short of recommended levels. The Introduction of oral supplements and enteral tube feeds results in increased energy and protein intake. Despite this, patients with a high macronutrient intake have low BMI levels, suggesting that subjects with a worse clinical status may have significantly higher nutrient needs. PS and post-transplant patients have a good clinical status and adequate BMI despite low energy intakes.

We recommend that dietary assessment is essential to identify the need for earlier nutritional intervention. The issue of how oral supplementation and enteral feeding can best meet additional energy requirements must be addressed. Diabetics, those on diet alone, PS and post-transplant patients must not be assessed by the mean of their population but by individual assessment. This must be annual with trusts funding adequate dietetic support and patients convinced of the need for complete assessment.

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