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Nutrient Status of Adults with Cystic Fibrosis

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Abstract

Nutrition is thought to influence disease status in patients with cystic fibrosis (CF). This cross-sectional study sought to evaluate nutrient intake and anthropometric data from 64 adult outpatients with cystic fibrosis. Nutrient intake from food and supplements was compared with the Dietary Reference Intakes for 16 nutrients and outcomes influenced by nutritional status. Attention was given to vitamin D and calcium given potential skeletal implications due to cystic fibrosis. Measurements included weight, height, body composition, pulmonary function, and serum metabolic parameters. Participants were interviewed about dietary intake, supplement use, pulmonary function, sunlight exposure, and pain. The participants' mean body mass index ($\pm$ standard deviation) was 21.8 ± 4.9 and pulmonary function tests were normal. Seventy-eight percent used pancreatic enzyme replacement for malabsorption. Vitamin D deficiency [25-hydroxyvitamin D (25OHD) < 37.5 nmol/L] was common: 25 (39%) were deficient despite adequate vitamin D intake. Lipid profiles were normal in the majority, even though total and saturated fat consumption represented 33.0% and 16.8% of energy intake, respectively. Reported protein intake represented 16.9% of total energy intake (range 10%–25%). For several nutrients, including vitamin D and calcium, intake from food and supplements in many participants exceeded recommended Tolerable Upper Intake Levels. Among adults with cystic fibrosis, vitamin D deficiency was common despite reported adequate intake, and lipid profiles were

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normal despite a relatively high fat intake. Mean protein consumption was adequate, but the range of intake was concerning, as both inadequate or excessive intake may have deleterious skeletal effects. These findings call into question the applicability of established nutrient thresholds for patients with cystic fibrosis.

Significant advances in the management of cystic fibrosis (CF) have increased the lifespan for patients with this common disease (1 in 3,900 white live births) (1). Previous research has suggested that patients with cystic fibrosis who have a good nutritional status have the best chance of maintaining and improving health (1–3). Nutritional therapy itself may also contribute to a better pulmonary status (4,5). A 1992 consensus report (1) emphasized the importance of detecting malnutrition in these patients to ensure optimal therapeutic management. A 2003 expert report on bone disease in cystic fibrosis cited both malnutrition and physical inactivity as risk factors for poor bone health (6).

Osteoporosis is a common health problem as patients with cystic fibrosis survive longer (6–11), and one study of patients with this disease showed that decreased bone density was more prevalent in the most malnourished patients (8). A subnormal body mass index (BMI), among other factors, can compromise bone density (6,11,12). A high fracture risk and kyphosis are also documented in these patients (12). Serum vitamin D concentrations may be an important predictor of skeletal health, but the relationship between vitamin D status and bone density remains unclear in this disease (6,9–15).

Most reports on nutrition in cystic fibrosis have been reviews, or include data collected in children (16–23). In this study, anthropometric, nutritional, and lifestyle variables were examined to determine their impact on health parameters in an outpatient sample of adult patients with cystic fibrosis, with the hypothesis that nutritional status would be compromised as reflected by a decreased intake of energy and specific nutrients, and low BMI and fat mass. The relationship between reported nutritional intake and Dietary Reference Intakes (DRIs) was examined, hypothesizing that vitamin D deficiency would be common and that many participants would not meet recommendations for this and other vitamins and nutrients. Among those with vitamin D deficiency, it was hypothesized that they would exhibit more skeletal problems and bone or generalized pain.

METHODS

Study Population

Sixty-four adult patients with cystic fibrosis who maintained stable weights were recruited from the Massachusetts General Hospital Cystic Fibrosis Care Center. Eligibility criteria included documentation of an elevated sweat chloride level or mutational analysis diagnostic for cystic fibrosis. From an original sample of 67 patients, three patients were excluded: two had undergone a lung transplant and one had a liver transplant.

Study Protocol

The Subcommittee on Human Studies of the Massachusetts General Hospital approved this cross-sectional study, and each subject gave informed consent. Data were obtained at the baseline visit of a prospective cohort study. A physical examination and medical interview were done, including questions about medical history, physical activity, menstrual and/or pubertal development, and food consumption. The history included questions about symptoms of malabsorption, and use of and general compliance with pancreatic enzyme replacement. Body composition was measured using skinfold thickness and bioelectrical impedance analysis. Subjects had fasting blood samples before 10:00 AM in the Mallinckrodt General Clinical Research Center at Massachusetts General Hospital.

Clinical Assessments

Anthropometric and Physical Examination Data—Height was measured by stadiometer (Holtain Limited, Crymych, UK). Subjects wore a hospital gown for weight measurements by digital scale (SR Instruments, Tonawanda, NY). BMI was calculated as kg/m^2 . Trained examiners used Lange calipers (Cambridge Scientific Industries, Cambridge, MA) to measure skinfolds three times at each site: triceps, biceps, subscapular, and suprailiac (24). Body fat was determined from age- and sex-adjusted tables (25). Estimation of percentage body fat composition was determined by tetrapolar bioelectric impedance analysis (BIA) (BIA 101A, RJL Systems, Inc, Clinton Township, MI) in the supine position (26,27). The mean mid-upper arm circumference was compared with mean for age of reference values (28).

Nutritional Intake—Two registered dietitians trained in food frequency questionnaire administration interviewed all subjects. Questions about food supplements and vitamin/mineral supplementation were included. Standard food models, cups, and measuring tools were used to evaluate intake. Nutrient calculations were done using the Nutrition Data System 2.93 software, developed by the Nutrition Coordinating Center, University of Minnesota, Minneapolis, MN (Food Database version 14, Nutrient Database version 29, released December 1998). Vitamin/mineral supplementation was assessed using the actual supplement label and verified with nutrient composition in the Nutrition Data System 2.93 software.

Metabolic/Pulmonary Function Data—Serum 25-hydroxyvitamin D (25OHD) and intact parathyroid hormone (PTH) concentrations were measured by radioimmunoassay (Diasorin, Stillwater, MN). All subjects completed simple lung spirometry within 1 month of the study visit.

Sun Exposure, Activity, and Pain—A sun exposure score was determined using a sunlight exposure questionnaire (29,30). The questionnaire asked subjects about time spent outdoors during the previous week and amount of skin exposure. A sunshine score was calculated: scores ranged from 0 to 9, with 9 indicating the highest exposure. Activity level was assessed using an interview-administered, self-report of activities during the previous 7 days (31). Light, moderate, hard, and very hard activity was calculated in hours per day. Questions about both generalized pain and bone pain were included in the interview.

Statistical Analysis

Descriptive statistics were generated for each variable. Pearson correlation analyses were calculated to determine the relationship between nutritional variables and specific lifestyle, functional, or anthropometric parameters. Significant predictors were then entered into multiple regression models to test significant relationships, with attention to potential confounding variables. Statistical analyses were done with SPSS Software (version 14.0, 2005, SPSS, Inc, Chicago, IL). All tests were two-tailed, and $P < 0.05$ was the significance threshold.

RESULTS AND DISCUSSION

The sample included 64 patients: 34 men and 30 women with cystic fibrosis, mean age ($\pm$ standard deviation) 31.4 ± 9.1 years. Sixty-two participants were white (97%) and 2 (3%) were black. The mean BMI for the sample was 21.8 ± 4.9 . The mean pubertal age for men was 13.9 ± 1.9 years (range 11 to 19), and mean age of menarche for women was 13.9 ± 2.4 years (range 8 to 19). Thirty-five participants (55.6%) had received oral glucocorticoid therapy, 6 (9%) had hepatobiliary disease, and 12 (19%) had cystic fibrosis-related diabetes

mellitus. Forty-nine (78%) were receiving pancreatic enzyme replacement therapy for malabsorption. Seventeen (27%) had kyphosis on physical examination, and 8 (12.5%) had scoliosis. Thirty-three (52%) had sustained one or more fractures.

The mean sunshine score was 5.4 ± 2.0 (range 0 to 9). The median level of light physical activity was 14.6 hours per day (range 7.9 to 17.3); 10 hours for moderate activity (range 0 to 48); 0.14 for hard exercise (range 0 to 21); and 0 for very hard exercise (range 0 to 20). Seventeen patients (27%) reported bone pain and 7 (11%) reported generalized pain.

Regarding nutritional intake, use of a combined vitamin A, D, E, and K preparation was reported by 56 participants (88%), whereas only 16 (25%) were consuming calcium supplements. Standard multivitamin supplements were used by 33 participants (52%). Intake of total energy, fat, carbohydrate, protein, vitamins, and minerals was assessed (Table 1). Based on weight, participants consumed 48.0 kcal/kg/day (54.6 kcal/kg/day for men vs 44.6 kcal/kg/day for women). The mean dietary percentages of fat, protein, and carbohydrate were determined. For total fat, the percentage was $33.0\% \pm 6.7\%$ (range 14 to 50), with the mean intake of saturated fat, $16.8\% \pm 7.3\%$ (range 4.5 to 35.8). For protein, the mean intake was $16.9\% \pm 2.9\%$ (range 10 to 24); and for carbohydrate, mean intake was $49.4\% \pm 8.6\%$ (range 14 to 68).

DRIs have been established for some nutrients (32) and are noted (Table 1). From diet and supplements, 57 patients (89%) were receiving Adequate Intake for calcium, with 20 patients (31%) exceeding the Tolerable Upper Intake Level (UL). Fifty-two (81%) patients were receiving the Adequate Intake for vitamin D and three participants (5%) exceeded the UL. Fifty-eight patients (91%) received the Recommended Dietary Allowance for magnesium, and none of the participants exceeded the UL. All participants met the Recommended Dietary Allowance for phosphorus, and five (8%) exceeded the UL. Nineteen patients (30%) exceeded the UL for folate, and one participant (1.5%) exceeded the UL for vitamin B-6. As a group, the mean intakes of magnesium, vitamin A, and niacin exceeded the UL (Table 1).

Twenty-five participants (39%) were found to be vitamin D-deficient ($25\text{OHD} < 37.5 \text{ nmol/L}$), replicating data from previous studies that showed a high prevalence of vitamin D deficiency among these patients (33–36). The deficiency was more common during winter: 12 cases identified with mean 25OHD for winter participants of $29.5 \pm 20.2 \text{ nmol/L}$ vs one case during summer with mean 25OHD for summer participants of $59.8 \pm 23.5 \text{ nmol/L}$ ($P < 0.001$). The correlation between 25OHD and PTH was not significant ($r = -0.11$, $P = \text{not significant}$), nor was the correlation between concentrations of either hormone and reported calcium or vitamin D intake. There was no correlation between sunshine score and serum concentrations of 25OHD , calcium, or PTH. Reported protein intake was positively correlated with intake of both calcium ($r = 0.70$, $P < 0.001$), vitamin D ($r = 0.59$, $P < 0.001$), and phosphorus ($r = 0.89$, $P < 0.001$), but was not significantly correlated with serum 25OHD , PTH, or calcium concentration. There were no differences with respect to BMI, energy intake, calcium intake, sun exposure, pulmonary function, or activity comparing those who were vitamin D-deficient vs not, nor was there a difference in vitamin D consumption (20.9 vs $24.2 \mu\text{g}$, respectively, $P = \text{not significant}$) between the groups. There was a significant correlation between 25OHD concentration and generalized pain ($r = -0.32$, $P = 0.016$), bone pain ($r = -0.31$, $P = 0.027$), and the presence of kyphosis ($r = -0.29$, $P = 0.035$). This seems to be the first report to suggest a relationship between vitamin D status and reported pain in patients with cystic fibrosis, replicating data from other patient groups (37).

The mean concentration of total cholesterol was normal, with levels less than 200 mg/dL (5.17 mmol/L) in 56 participants (88%) (Table 2). Similarly, the mean triglyceride level was

normal. Consistent with recommendations from cystic fibrosis providers about liberalizing fat (1,3,22,38), this cohort consumed a substantial intake of total and saturated fat, yet serum lipid levels were normal. Pencharz and Durie also found that a high-energy, high-fat diet in adults with cystic fibrosis did not increase serum lipid levels in patients with pancreatic insufficiency (38); this is of relevance to the current study in which the majority of participants exhibited malabsorption. Other studies have shown lower mean total cholesterol levels in patients with cystic fibrosis (39,40). With the exception of those few patients with pancreatic sufficiency in whom a long-term, high-fat intake may be associated with increased cardiovascular risk, the current recommendation for a high-fat diet (38,41) does not seem to result in an unfavorable metabolic profile in these patients.

Adequate protein seems to confer skeletal benefits, whereas excessive amounts may lead to hypercalciuria (42). In the current sample, the mean reported protein intake suggested adequate overall consumption, but the wide range of intake was concerning. In some participants, protein intake exceeded the recommended threshold, which could potentially lead to hypercalciuria and skeletal losses; inadequate consumption in others could compromise both bone mass and strength (42). Protein intake did not seem to affect calcium accretion as there was no significant relationship found between protein intake and circulating levels of calcium or vitamin D. There was a positive correlation noted between protein intake and calcium, phosphorus, and vitamin D intakes, which may reflect the participants' consumption of these nutrients through dairy products.

The study sample was a well-nourished cohort, reflected by mean BMI and percentage body fat, and normal serum levels of albumin and total protein (Table 2). There were sex differences noted for mid-upper arm circumference: the mean for the male participants was between the 5th and 10th percentile of reference values for healthy men, and for females was between the 25th and 50th percentile for healthy women (28). Interestingly, there were no significant intake or BMI differences by sex to explain the dimorphic trends observed. There were no correlations noted between serum albumin and total protein and BMI, or reported consumption of total energy, protein, fat, or carbohydrate.

Pulmonary function test results are shown in Table 2; they were not significantly correlated with BMI, total energy intake, or protein intake. There was a modest positive trend between forced expiratory capacity in one second and energy intake ($r=0.25$, $P=0.066$).

Limitations of this study should be acknowledged. The sample size was small, which limited statistical power to examine secondary questions of interest outside of the primary aims; other hypotheses were not tested because of the small size of subgroups. Because the participants were recruited from a tertiary care center, selection bias could have been introduced. However, patients were recruited at their baseline level of health, and three patients who had received transplants were excluded because they were substantially malnourished. Trained dietitians obtained dietary intake data using validated assessment tools, but data were still vulnerable to the limitations of self-report. The current data were obtained from a sample among which the majority of participants were receiving pancreatic enzyme replacement, and the results generated are thus applicable primarily to those patients with malabsorption. Detailed data about compliance with this therapy were not obtained. Urinary calcium concentrations also were not obtained and would have been helpful in evaluating whether calcium absorption was altered by protein intake. Lastly, although this report provides intake data for specific nutrients and compares them with the DRI, there are no outcomes data reported (eg, bone density data), which limits the ability to confirm whether dietary intake is adequate in these patients. However, this comparison represents a starting point in describing the typical intake of nutrients in patients with cystic fibrosis. These preliminary findings will hopefully stimulate further research in this area.

CONCLUSIONS

This study seems to be the first in adult patients with cystic fibrosis to examine reported nutrient intake with respect to the DRIs. Although saturated fat consumption represented 16.8% of daily energy intake, lipid profiles were normal in the majority of participants, supporting the current common practice to liberalize fat in the diet of these patients. The mean protein consumption was normal, but the range of intake was concerning, especially with respect to bone health. Lastly, these data emphasize that vitamin D deficiency is common among these patients, with the potential to negatively impact their functional status.

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Table 1

Nutritional intake (food and supplements) of participants with cystic fibrosis

Nutritional variable	Food and supplement intake mean±SD ^c	Range	DRI ^a	UL ^b
Calories (kcal/d)	2,962.1±1,180.8	1,084.1–7,301.5	120%–150% ^d	ND ^e
Total fat (g/d)	123.8±91.4	26.8–592.6	<i>f</i>	ND
Saturated (g/d)	49.1±32.3	11.9–187	<i>f</i>	ND
Monounsaturated (g/d)	42.3±27.3	12.7–162.5	—	ND
Polyunsaturated (g/d)	17.3±13.9	4.8–94.8	—	ND
Carbohydrate (g/d)	366.7±145.1	135.1–955.6	130	ND
Protein (g/d)	126.3±53.7	41.4–282.3	56-M 46-F	ND
Calcium (mg)	2,098±1,134	426–6,145	1,000 M ^g 1,200 F ^g	2,500
Vitamin D (μg)	23.2±12.6	1.9–55.9	5 ^g	50
Magnesium (mg)	563±286	169–1,779	420-M 310-F	350
Phosphorus (mg)	2,284±1,306	728–7,550	700	4,000
Copper (μg)	3,400±1,900	900–8,500	900	10,000
Vitamin C (mg)	359.0±362.8	48.5–1,731.8	90-M 75-F	2,000
Selenium (μg)	360.1±1,339.9	60.6–10,265.3	55	400
Vitamin A (μg RAE) ^h	3,706±2,108	342–10,266	900-M 700-F	3,000
Beta-carotene (μg)	7,120±4,688	1553–22,460	ND	ND
Thiamin (mg)	6.8±13.5	1.2–105.5	1.2-M 1.1-F	ND
Riboflavin (mg)	9.7±15.7	2–106.8	1.3-M 1.1-F	ND
Niacin (mg)	58.9±28.9	21.1–170.6	16-M 14-F	35
Folate (μg) ⁱ	854±376	200–1,849	400	1,000
Vitamin B6 (mg)	7.7±13.6	1.2–36.9	1.3	100
Vitamin B12 (μg)	22.5±24.5	3.9–1,555.5	2.4	ND
Zinc (mg)	31.1±15.9	6.5–91.6	11-M 8-F	40

^aDRIs=Dietary Reference Intakes for adults age 19–30 years (reference 32). M=male. F=female.^bUL=tolerable upper intake level.^cSD=standard deviation.^d100%–150% of the Estimated Energy Requirement for healthy children and adults (references 16–18,20).^eND=not determined.^f<25%–35% of total calories from fat, with <10% from saturated fat.^gAdequate Intakes (AIs) are footnoted, whereas Recommended Dietary Allowances (RDAs) are not.^hRAE=retinol activity equivalent; 1 RAE=1 μg retinol.

ⁱ As dietary folate equivalents (DFE). 1 DFE=1 μg food folate=0.6 μg of folic acid from fortified food or as a supplement consumed with food=0.5 μg of a supplement taken on an empty stomach.

Table 2

Biochemical parameters and pulmonary function results for 64 adults with cystic fibrosis

	Mean±SD ^a (range)	Normal values
Metabolic studies		
PTH ^b (pg/mL)	40.4±28.5 (8–163)	10–65
25(OH)D ^c (ng/mL)	19.4±13.1 (8–68)	10–55
Calcium (mg/dL) ^d	9.1±0.44 (8.1–10)	8.0–10.5
Phosphorus (mg/dL) ^e	3.4±0.62 (2.2–4.9)	2.7–4.9
Magnesium (mg/dL) ^f	1.7±0.24 (1.3–2.3)	1.5–2.2
Total cholesterol (mg/dL) ^g	145.6±44.2 (70–295)	Optimal <200
Triglycerides (mg/dL) ^h	114.7±71.5	Optimal <250
Total protein (g/dL) ⁱ	7.3±0.55 (6–8.7)	5.5–8.2
Albumin (g/dL) ^j	4.0±0.43 (3.1–4.8)	3.0–4.6
Pulmonary function tests		
FEV1 ^k (% predicted)	56.7±24.1 (23–106)	
FVC ^l (% predicted)	72.7±21.2 (35–111)	
Body composition data		
MUAC ^m (mm)	27.1±43.6 (20.6–36.6)	
% Body fat (BIA) ⁿ	21.0±7.2 (6–37)	
% Body fat (skinfolds)	20.5±7.7 (4.8–36.8)	

^aSD=standard deviation.^bPTH=parathyroid hormone. To convert pg/mL PTH to ng/L, multiply pg/mL by 1.0.^c25(OH)D=25-hydroxyvitamin D. To convert ng/mL 25(OH)D to nmol/L, multiply ng/mL by 2.496.^dTo convert mg/dL calcium to mmol/L, multiply mg/dL by 0.25.^eTo convert mg/dL phosphorus to mmol/L, multiply mg/dL by 0.323.^fTo convert mg/dL magnesium to mmol/L, multiply mg/dL by 0.411.^gTo convert mg/dL cholesterol to mmol/L, multiply mg/dL by 0.0259.^hTo convert mg/dL triglycerides to mmol/L, multiply mg/dL by 0.0113.ⁱTo convert g/dL total protein to g/L, multiply g/dL by 10.0.^jTo convert g/dL albumin to g/L, multiply g/dL by 10.0.^kFEV1=forced expiratory volume in first second.^lFVC=forced vital capacity.^mMUAC=mid-upper arm circumference.ⁿBIA=bioelectrical impedance analysis.