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Original Article

Dietary intakes and quality of Irish adults with cystic fibrosis: Comparisons to nutrition guidelines and HEI-2020

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ABSTRACT

Background: With evolving Cystic Fibrosis (CF) phenotypes resulting from changes to clinical management, healthier dietary practices are warranted for many people with CF. Whilst diet composition is reported, diet quality data is lacking in CF. This study aims to evaluate dietary intakes and diet quality in adults with CF via guideline comparison and a validated diet quality index.

Methods: Cross-sectional study of Irish adults with CF. Demographic questionnaires and three-day food diaries were completed. Healthy Eating Index – 2020 (HEI-2020) assessed diet quality. Data was statistically analysed in SPSS®.

Results: Of $n = 68$ participants (female: 58.8 %, age: 35.2 ± 10.1 years, FEV_1%: 77.4 ± 25.1 %), 36.8 % were overweight/obese and 77.6 % pancreatic insufficient. While median (interquartile range) percentage estimated average requirement (EAR) was 110.1 (45.3) %, 50.0 % of participants were below CF energy requirements (110 % EAR). Mean percentage total energy intake (%TEI) protein (18.0 ± 3.9 %) aligned to dietary reference values (DRV). %TEI carbohydrates (44.1 ± 6.5 %) was below, and %TEI fat (37.1 ± 5.4 %), saturated fat (14.1 ± 3.3 %) and sugar (17.4 ± 5.6 %) exceeded DRV. Median vitamin A intake was adequate [936.6 (1005.2) μg], but vitamin D [3.6 (4.3) μg], E (9.6 ± 5.2 mg) and K_1 [31.1 (71.2) μg] intakes were insufficient without supplementation.

Regarding Irish healthy eating guidelines, 95.6 % of participants overconsumed energy-dense nutrient poor (EDNP) foods, with 76.5 % below vegetables, salad and fruit intake guidelines. Participants' mean HEI-2020 score (0–100) was 59.3 ± 12.4.

Conclusion: Findings indicate suboptimal diet quality. Despite reliance on EDNP foods, many did not achieve energy targets. Moving forward, emphasis on diet quality is of paramount importance to improve overall health in people with CF.

Abbreviations: CF, Cystic Fibrosis; PwCF, People Living With Cystic Fibrosis; PI, Pancreatic Insufficiency; VSTs, Variant Specific Therapies; EAR, Estimated Average Requirement; BMI, Body Mass Index; EDNP, Energy-Dense Nutrient-Poor; HEI-2020, Healthy Eating Index 2020; SD, Standard Deviation; FEV_1%, Predicted Percentage Forced Expiratory Volume; %TEI, Percentage Of Total Energy Intake; DRV, Dietary Reference Values.

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1. Introduction

Cystic Fibrosis (CF) has evolved considerably over the last decade with significant improvements in outcomes [1] directly related to advancements in variant specific therapies (VSTs) and better clinical care and multidisciplinary management of CF [2]. This has extended the median survival age of people living with CF (PwCF) to above 50 years old in many countries including Canada, USA, Australia, New Zealand, and some European countries[3,4].

Pulmonary infections, increased resting energy expenditure from respiratory effort, and food and nutrient malabsorption have typically resulted in increased energy requirements for PwCF [5]. International nutrient guidelines for PwCF have reflected this in consistently recommending increased energy intakes relative to estimated average requirements (EAR) [USA (preceding 2020) / Europe / Australia: 110–200 % EAR [6–8]; USA (2020): 125 % EAR [9]]. Until recent years international guidelines recommended a high energy, high fat diet with pancreatic enzyme replacement therapy, to meet body mass index (BMI) targets [6,8,10]. However, in 2020 up to date guidelines from the Academy of Nutrition and Dietetics suggest high fat diets are not therapeutic beyond meeting high energy targets, and PwCF should follow general population macronutrient guidelines [9]. Likewise, the latest European guidelines (2024) state that insufficient evidence is available to advise deviation from national recommendations on macronutrient balance [7].

Unfavourable dietary patterns are potentially placing PwCF at risk of developing cardiovascular disease, obesity and other morbidities associated with a poorer diet quality [11,12] in combination with VSTs. The most recent data registry report (2023) from the CF Registry of Ireland indicated that 43.6 % of Irish adult PwCF are overweight or obese [13] and increases in overweight / obesity have been attributed to the introduction of VSTs [14]. Research assessing weight and body compositional change and the use of ivacaftor [15–17], lumacaftor-ivacaftor [18,19], tezacaftor-ivacaftor [19] and ivacaftor-tezacaftor-elexacaftor [20–22] support this relationship. However, the increases in weight and BMI are likely through a combination of factors, including mitigation of malabsorption and other metabolic processes that previously limited weight gain prior to VSTs, and the continuation of high caloric intakes [17]. A higher risk of major cardiac disease events have also been reported for adult PwCF compared to matched controls (hazard ratio: 1.75, 95 % Confidence Interval: 1.59–1.92; $p < 0.001$) [23].

While historical dietary practices remain relevant for some PwCF, many should be encouraged to follow a healthier dietary pattern which focuses on diet quality as well as quantity [7]. Comparisons to national dietary guidelines can serve as a means of assessing diet quality, but alternatively, various metric tools have been developed to evaluate diet quality [24–26], providing a means of assessing and mapping overall diet quality to objective measures [27]. A previous systematic review highlighted a paucity of diet quality research amongst adult PwCF ($n = 4$ studies) and where reported, diet quality was suboptimal with a lack of heterogeneity in measurement tools [28].

In a modulator era, the disease process has rapidly changed and the impact of diet on underlying pathogenic mechanisms is largely unknown. However, it is well established that diet quality can impact inflammatory processes in other diseases [29–31]. Furthermore, with increases in life expectancy PwCF are becoming more at risk of developing diet-related chronic diseases[32,33]. As such, subjective and objective dietary measures need to be assessed and considered in accelerating metabolic disease processes, similar to the non-CF population. With few studies investigating diet quality thoroughly and no research to date reporting Irish dietary intakes and patterns in adult PwCF in a post modulator era, this study will endeavour to provide a comprehensive evaluation of the dietary intakes and diet quality of Irish adult PwCF.

2. Methods

2.1. Study design and population

This observational, cross-sectional study is reported in accordance with STROBE guidelines [34]. Quantitative data were collected using demographic and self-reported health related questionnaires and an estimated three-day food diary (September 2020 - March 2023). Study material was available in digital and paper formats. Questionnaire data were collected and managed via the Electronic Data Capture system, Castor, Version 1.6 (Ciwit B.V., The Netherlands). The smartphone application, Libro, from the dietary analysis software, Nutritics, Version 5.7 Research Edition (Nutritics Ltd.: Dublin, Ireland) [35] was used to gather three-day food diary data digitally. The study protocol [36] was approved by the University of Limerick Institutional Review Board and relevant hospital ethics committees, with all participants providing written informed consent.

2.1.1. Participant recruitment

The aim was to recruit between 70–100 adult PwCF using a convenience sampling method [37]. Recruitment was carried out through CF Ireland (patient advocacy organisation) membership forums and in CF centres / clinics. Dietitians initially forwarded a recruitment email to the centres / clinics mailing lists. Additionally, dietitians actively promoted the study at centre / clinic visits. One centre / clinic adopted a different approach due to a large number of PwCF attending. The dietitian actively pre-screened participants prior to contact via email or at centre / clinic visits. All other participants were screened using a self-report questionnaire created in Microsoft Forms specifically designed to assess eligibility based on inclusion / exclusion criteria.

2.1.2. Participant eligibility

Inclusion criteria: Adults (≥18 years old) with a diagnosis of CF, living in Ireland. *Exclusion criteria:* Receiving enteral nutrition, following a prescription diet for another medical condition (e.g., coeliac disease), were pregnant or has previously received an organ-transplant. Adults must have been on a stable medical regimen for at least four weeks prior to commencing the study with no recent pulmonary exacerbations or administration of oral or intravenous antibiotics or glucocorticoids.

2.2. Assessment of dietary intake

Three-day food diary logs were used to evaluate dietary intakes and compliance to dietary guidelines. Once consent was obtained, participants were instructed to use the Libro app to record all foods and beverages consumed over a three-day period. Although weighing of foods and fluids was not required, participants were asked to estimate portion sizes using common household measurements and to document the cooking methods used. Data were automatically exported to Nutritics for analysis, generating comprehensive daily and mean energy, macronutrient, and micronutrient intake values using Nutritics, Ireland (2009 Irish Food Composition Database). Food diaries underwent data quality integrity checks, ensuring that any implausible values (i.e., extreme calorie intakes or unrealistic food portions) were identified and addressed. Oral nutritional supplements were included in food logs as they can significantly contribute to energy and nutrient intakes [38]. Although the use of micronutrient supplements are commonplace in PwCF [7], contributions from micronutrient supplements were not included in the final dietary intake reports, to examine the sufficiency of micronutrient intakes through diet alone. Dietary intakes were evaluated by assessing adherence to appropriate CF-specific [7] and general population nutrition guidelines [39–42].

2.3. Calculating diet quality scores

The Healthy Eating Index – 2020 (HEI-2020) tool was used to

evaluate diet quality (Appendix 1). Although developed for the American context, the HEI-2020 is a valuable tool for assessing diet quality, as it captures core nutritional components relevant to the Irish population, with its principles aligning with Irish dietary guidelines (i.e., promoting fruits, vegetables, whole grains, and lean proteins) [42], and previous versions have been effectively used to examine diet–health relationships in Irish cohorts [43,44]. Macronutrient and micronutrient intakes were quantified per 1000 kcals. Grams of food items were converted to appropriate serving equivalents (i.e., cup, ounce) using the Food Pattern Equivalent Database [45]. Thereafter, the simple Healthy Eating Index scoring algorithm was applied [46] and HEI-2020 scores were calculated ranging from 0–100, with higher scores indicating higher diet quality. A score between 0–59 is the lowest grade applicable to HEI-2020 indicating poor diet quality [26].

2.4. Statistical analysis

Statistical analysis was conducted with SPSS® Statistics for Windows, Version 29 (IBM Corp., Released 2022). Macronutrient and micronutrient intakes were quantified as absolute intake. Descriptive statistics were used to describe baseline characteristics of the cohort. Distribution of data was tested using the Kolmogorov Smirnov test of normality, where a significance of $p > 0.05$ identified data as normally distributed. Data were presented as means ± standard deviations, median (interquartile range) and frequencies (*n* and %) where appropriate. Where variables were not normally distributed, variables were noted with an asterisk. Parametric and non-parametric tests (e.g., *t*-tests, ANOVA, Mann-Whitney U, Kruskal-Wallis) were applied to compare continuous variables, with post hoc tests used for specific comparisons where necessary. Pearson's Chi-square was used to test differences between categorical variables. Total diet quality scores and scores for each component of the score were calculated and compared between demographic groups using appropriate statistical tests as previously described. Significance is denoted as $p < 0.05$.

3. Results

3.1. Demographic and clinical characteristics

The demographic and clinical characteristics of participants are displayed in Table 1. Of $N = 73$ enrollees to the study, $n = 1$ participant submitted the demographic and self-reported health questionnaire partially completed. A total of $n = 68$ three-day food diaries were completed as part of this study. Reasons for food diaries being excluded included participants not returning the food diaries after three requests ($n = 4$), or food diaries being returned with insufficient information provided ($n = 1$). This resulted in $n = 68$ HEI-2020 scores being generated.

The mean age of participants was 35.2 ± 10.1 years (female: 58.8 %). The median BMI was 24.3 ± 4.2 kg/m^2 with male participants having a significantly higher BMI compared to female participants ($p < 0.001$). The proportion of individuals classified as overweight or obese (World Health Organisation cut-off: ≥25 kg/m^2) [47] was 36.8 %, with significantly more male participants (57.1 %) classified as overweight or obese (female: 22.5 %, $p = 0.027$). A total of 77.6 % of participants reported pancreatic insufficiency (PI) and 28.4 % reported CF-related diabetes. The mean predicted percentage forced expiratory volume (FEV_1%) was 77.4 ± 25.1 % and 80.6 % of participants reported taking modulators at the time of the study.

3.2. Dietary intakes

3.2.1. Energy, macronutrient, and micronutrient intakes

The energy, macronutrient, micronutrient, and food serve intakes for the cohort are displayed in Table 2. The median energy intake for participants was 2281.8 (906.7) kcals/day and median percentage EAR was

Table 1
Demographic and clinical characteristics of Irish adults living with CF who participated in this cross-sectional study.

	All (*N* = 68)	Male (*n* = 28)	Female (*n* = 40)	*p*-value
Age (y) (*range: 19–68*)	35.2 ± 10.1	37.3 ± 10.5	33.7 ± 9.7	0.150
BMI (kg/m^2)	23.5 (3.9)	25.3 (4.2)	22.3 (3.4)	<0.001
BMI Classification *n* (%)[*,~]				0.027
Underweight (<20)	1 (1.5)	0 (0.0)	1 (2.5)	–
Normal (20–24.9)	42 (61.8)	12 (42.9)	30 (75.0)	–
Overweight (25–29.9)	21 (30.9)	14 (50.0)	7 (17.5)	–
Obese (≥30)	4 (5.9)	2 (7.1)	2 (5.0)	–
	***n* = 67**		***n* = 39**	
FEV_1%	77.4 ± 25.1	76.5 ± 26.7	78.1 ± 24.2	0.801
FEV_1% Categories *n*(%)				0.880
<40 %	6 (9.0)	3 (10.7)	3 (7.7)	–
40–69.99 %	17 (25.4)	8 (28.6)	9 (23.1)	–
70–79.99 %	9 (13.4)	3 (10.7)	6 (15.4)	–
≥80 %	35 (52.2)	14 (50.0)	2 (53.8)	–
Hospital Admission				–
Since last admission (days)[*]	832 (1093.0)	985.0 (952.8)	773.0 (1143.0)	0.247
Last admission length (days)[*]	14.0 (9.0)	13.0 (8.5)	14.0 (10.0)	0.964
Admitted in last year n(%)	48 (70.6)	23 (82.1)	25 (64.1)	0.106
Highest Education *n*(%)				0.507
Secondary level	9 (13.4)	3 (10.7)	6 (15.4)	–
Third level[‡]	39 (58.2)	15 (53.6)	24 (61.5)	–
Postgraduate level	19 (28.4)	10 (35.7)	9 (23.1)	–
Income Status *n*(%)				0.003
Full income	29 (43.3)	19 (67.9)	10 (25.6)	–
Part-time income	13 (19.4)	3 (10.7)	10 (25.6)	–
No work-based income	25 (37.3)	6 (21.4)	19 (48.7)	–
Area of Living *n*(%)				0.061
City	27 (40.3)	15 (53.6)	12 (30.8)	–
Town	11 (16.4)	6 (21.4)	5 (12.8)	–
Village	8 (11.9)	3 (10.7)	5 (12.8)	–
Countryside	21 (31.3)	4 (14.3)	17 (43.6)	–
Living Situation *n*(%)				0.512
Living alone	7 (10.4)	4 (14.3)	3 (7.7)	–
Living with housemates	13 (19.4)	4 (14.3)	9 (23.1)	–
Living with family/relatives	47 (70.1)	20 (71.4)	27 (69.2)	–
Clinical Characteristics *n*(%)				
Pancreatic insufficiency	48 (71.6)	22 (78.6)	26 (66.7)	0.286
CF-related diabetes	19 (28.4)	9 (32.1)	10 (25.6)	0.790
GORD	16 (23.5)	7 (25.0)	9 (23.1)	0.856
CF-related liver disease	8 (11.8)	6 (21.4)	2 (5.1)	0.050
CF-related bone disease	6 (9.0)	1 (3.6)	5 (12.8)	0.191
Medication / Supplement Use *n*(%)[†]				
VSTs	54 (80.6)	20 (71.4)	34 (87.2)	0.108
Antibiotics	35 (52.2)	17 (60.7)	18 (46.2)	0.239
Mucolytics	13 (19.4)	6 (21.4)	7 (17.9)	0.722
Bronchodilators	27 (40.3)	12 (42.9)	15 (38.5)	0.718
Insulin	19 (28.4)	9 (32.1)	10 (25.6)	0.560
Steroids	10 (14.9)	7 (25.0)	3 (7.7)	0.054
Fat-soluble vitamins	57 (85.1)	22 (78.6)	35 (89.7)	0.179
Oral Nutritional Supplements	10 (14.7)	6 (21.4)	4 (10.3)	0.179

Parametric variables are presented as mean ± SD and non-parametric variables as median (interquartile range).
P-values presented were derived with chi-square tests, independent sample *t*-tests and ANOVA tests (Non-parametric: Mann Whitney U and Kruskal-Wallis tests).
Significance derived from a *p*-value of <0.05.
[*] Non-parametric test.
[~] World Health Organisation cut-off values [47].
[†] Chronic medications and supplements used on a regular basis.
[‡] University, college or vocational education.
Abbreviations: BMI, body mass index; FEV_1%, percentage of predicted force expiratory volume for age, sex, ethnicity, and height; GORD, gastro-oesophageal reflux disease; VSTs, variant-specific therapies; SD, standard deviation.

Table 2
Energy, macronutrient, micronutrient, and food serve intakes of Irish adults with CF who participated in this cross-sectional study.

Nutrient Measure	All ($n = 68$)	Male ($n = 28$)	Female ($n = 40$)	*p*-value	Recommended Intakes	
					Male	*Female*
Energy (kcal)*	2281.8 (906.7)	2456.7 (1122.5)	2157.0 (788.8)	0.050	2200–5000[a]	1980–4000[a]
% EAR[*,†]	110.1 (45.3)	109.2 (49.9)	113.5 (41.5)	0.770	110–200[a]	
Carbohydrates (g)*	248.5 (101.0)	256.3 (102.8)	236.9 (78.0)	0.105	-	
%TEI carbohydrates	44.1 ± 6.5	43.9 ± 7.1	44.3 ± 6.1	0.814	45–60	
Protein (g)	107.2 ± 36.4	117.9 ± 33.4	99.6 ± 36.8	0.040	-	
%TEI protein	18.0 ± 3.9	18.8 ± 4.5	17.5 ± 3.4	0.167	15–20[b]	
Fat (g)	100.4 ± 37.2	103.2 ± 28.0	98.5 ± 42.6	0.610	-	
%TEI fat	37.1 ± 5.4	36.3 ± 5.2	37.7 ± 5.5	0.302	20–35	
Fibre (g)	23.0 ± 9.0	22.8 ± 7.9	23.0 ± 9.8	0.924	25	
Total sugar (g)*	98.2 (57.05)	102.3 (59.7)	97.3 (60.6)	0.462	as low as possible	
%TEI total sugar	17.4 ± 5.6	17.2 ± 5.5	17.5 ± 5.8	0.799	<10[c]	
Free sugar (g)*,~	33.8 (36.7)	43.0 (25.8)	29.4 (42.4)	0.079	as low as possible	
*%TEI free sugar**	6.0 (4.2)	6.9 (3.4)	4.7 (6.0)	0.121	<5[d]	
Saturated fats (g)	38.1 ± 15.8	38.7 ± 12.5	37.7 ± 17.9	0.804	as low as possible	
%TEI saturated fat	14.1 ± 3.3	13.6 ± 3.2	14.4 ± 3.4	0.365	<10[c]	
Monounsaturated fat (g)	31.3 ± 15.8	33.7 ± 13.7	29.6 ± 17.0	0.296	–	
%Fat Monounsaturated	30.4 ± 8.0	31.2 ± 7.9	29.1 ± 7.9	0.905	–	
Polyunsaturated fat (g)	12.1 ± 6.3	12.2 ± 5.2	12.0 ± 7.1	0.113	–	
%Fat Polyunsaturated	11.8 ± 3.8	11.7 ± 3.4	11.8 ± 4.1	0.874	–	
Omega 3 (n-3) (g)*	0.96 (1.5)	1.0 (1.3)	0.9 (1.6)	0.695	–	
Omega 6 (n-6) (g)*	4.7 (6.1)	5.9 (5.0)	4.2 (6.2)	0.363	–	
Sodium (mg)*	2657.4 (1256.6)	2717.5 (1409.8)	2650.2 (1287.0)	0.737	2000[e]	
Calcium (mg)	1066.7 ± 494.6	1201.9 ± 465.0	972.1 ± 498.3	0.059	18–25 years: 1000 >25 years: 950	
Iron (mg)*	11.3 (7.9)	13.6 (7.0)	9.3 (6.9)	0.095	11	16
Zinc (mg)	11.0 ± 4.9	12.6 ± 4.9	9.8 ± 4.6	0.019	9.4–16.3	7.5–12.5
Vitamin A (ret. eq.) (μg)*	936.6 (1005.2)	846.8 (1189.7)	943.7 (951.7)	0.891	750[f]	650[f]
Vitamin D (μg)*	3.6 (4.3)	4.0 (4.8)	3.6 (3.6)	0.751	15[f]	
Vitamin E (mg)	9.6 ± 5.2	9.9 ± 4.2	9.4 ± 5.8	0.667	13[f]	11[f]
Vitamin K_1 (μg)*	31.1 (71.2)	31.0 (77.3)	31.1 (65.1)	0.955	70[f]	
Vitamin C (mg)*	87.5 (98.4)	104.1 (123.0)	84.5 (98.9)	0.370	110	95
Food Serves (serves / day)[g]						
Vegetables, salad & fruit	3.5 ± 2.5	3.4 ± 2.2	3.6 ± 2.6	0.695	5–7	
Grains[‡]	3.6 ± 1.3	3.9 ± 1.4	3.4 ± 1.1	0.134	3–5[h]	
Milk, yoghurt & cheese*	2.7 (2.3)	2.7 (2.6)	2.7 (2.2)	0.818	3	
Protein foods*,[§]	3.0 (2.7)	3.3 (2.1)	2.8 (1.6)	0.101	2	
Fats, spreads & oils	4.1 ± 2.9	4.0 ± 3.0	4.2 ± 2.8	0.763	very small amounts	
EDNP foods*,[‖]	5.1 (4.7)	5.2 (4.6)	4.5 (4.6)	0.485	not every day	

Parametric variables are presented as mean ± SD and non-parametric variables as median (interquartile range).
P-values were derived with independent sample *t*-tests (Non-parametric: Mann Whitney *U* test).
Significance derived from a *p*-value of <0.05.
Guideline intakes are EFSA general population dietary reference values [39] unless stated otherwise.
Calcium, iron, zinc, vitamin A and C recommended intakes are derived from EFSA population reference intake values.
Vitamins A, E, and K recommended intakes are derived from EFSA adequate intake values.

* Non-parametric test.

~ Free sugars refer to sugars added to foods as well as sugars naturally present in honey, syrups, and fruit juices. It excludes sugars found in whole fruits and vegetables [40].

† Derived from the average energy intake recommendations (female: 1800–2000 kcal/day, male: 2000–2500 kcal/day) provided by Healthy Ireland [42].

‡ Wholemeal cereals and breads, potatoes, pasta, and rice.

§ Meat, poultry, fish, eggs, beans and nuts.

‖ Foods and drinks high in fat, sugar and salt.

[a] Derived from ESPEN-ESPGHAN-ECFS CF-specific nutrition guidelines [7].

[b] Derived from Academy of Nutrition and Dietetics CF-specific nutrition guidelines [9].

[c] Derived from the Irish Heart Foundations nutrition guidelines for heart health [41].

[d] Derived from World Health Organisation nutrition guidelines [40].

[e] CF-specific nutrition guidelines recommend supplementing in stress situations when excessive sweating is expected (i.e., fever, exercise/sports, hot weather) [7].

[f] CF-specific nutrition guidelines recommend to always take a supplement on top of recommended intakes [7].

[g] Derived from Healthy Ireland guidelines [42].

[h] Up to 7 serves per day for men 19–50 years old.

Abbreviations: % EAR, percentage estimated average requirement; %TEI, percentage of total energy intake from; ret. eq., retinol equivalent; SD, standard deviation; EDNP, Energy-Dense Nutrient Poor; EFSA, European Food Safety Authority; CF, Cystic Fibrosis.

reported as 110.1 (45.3) %, with 50.0 % of participants not achieving the minimum requirement for PwCF (110 % EAR) [7] (Appendix 2). The mean percentage of total energy intake (%TEI) from carbohydrates and protein were reported as 44.1 ± 6.5 % and 18.0 ± 3.9 %, respectively. A total of 72.1 % of participants were above the general population dietary reference value (DRV) for %TEI fat (20–35 %TEI) [39] (Appendix 2).

The mean %TEI total sugar and saturated fat were above heart health guidelines [41] and fibre intakes were insufficient relative to general population DRV (25 g/day), as were intakes (without micronutrient supplementation) for fat-soluble vitamins D, E, and K_1 [39]. Vitamin A and calcium intakes were sufficient relative to DRVs [39]. Furthermore, the median intake of iron was suboptimal in female participants relative to gender-specific guidelines (16 μg), with the median iron intake for male participants meeting the reference intake value (11 μg) [39].

3.2.2. Food serve intakes

In relation to food groups, mean serves of vegetables, salad and fruit were suboptimal with 76.5 % of participants not meeting the Irish food based dietary guidelines [42]. Mean serves of fats, spreads and oils, and median serves of protein foods and EDNP foods exceeded guidelines with 80.9 % of participants overconsuming fats spreads and oils, 67.6 % overconsuming protein foods, and 95.6 % overconsuming EDNP foods (Appendix 2) [42]. Compared to Irish food based dietary guidelines [42], mean serves of grains and median serves of milk, yoghurt and cheese were sufficient.

3.3. Diet quality scores

The mean HEI-2020 score was 59.3 ± 12.4. Participants with a FEV_1% above the mean value had significantly higher HEI-2020 diet quality scores (62.8 ± 11.2, $p = 0.009$) and total fruits [3.0 (3.0), $p = 0.017$] and whole fruits [4.5 (4.0), $p = 0.025$] component scores compared to participants below the mean FEV_1% [HEI-2020: 54.8 ± 12.9; total fruits: 1.0 (1.0); whole fruits: 1.0 (3.0)] (Table 3). Further subgroup analysis (i.e., gender, FEV_1%, BMI classification, PI status, modulator use) of dietary intakes, food serves and diet quality can be found in Appendix 3.

Table 3
HEI-2020 diet quality score and associated component scores in Irish adults with CF who participated in this cross-sectional study below and above the mean FEV_1% for the cohort.

Diet Quality Measure	All ($n = 68$)	Below Mean FEV_1% ($n = 29$)	Above Mean FEV_1% ($n = 38$)	*p*-value
HEI-2020 Score (0–100)	59.3 ± 12.4	54.8 ± 12.9	62.8 ± 11.2	0.009
HEI-2020 Component Scores*				
Total Fruits (0–5)	2.0 (2.8)	1.0 (1.0)	3.0 (3.0)	0.017
Whole Fruits (0–5)	3.0 (4.0)	1.0 (3.0)	4.5 (4.0)	0.025
Total Vegetables (0–5)	4.0 (2.0)	4.0 (2.0)	5.0 (2.0)	0.167
Greens & Beans (0–5)	3.0 (3.8)	2.0 (4.5)	3.5 (3.0)	0.131
Wholegrains (0–10)	5.0 (5.0)	4.0 (6.5)	5.0 (4.3)	0.665
Dairy (0–10)	6.0 (5.0)	5.0 (4.5)	7.0 (6.0)	0.206
Total Protein (0–5)	5.0 (1.0)	5.0 (1.0)	5.0 (1.0)	0.475
Seafood & Plant Protein (0–5)	3.0 (5.0)	3.0 (5.0)	3.5 (3.3)	0.163
Refined Grains (0–10)	8.0 (4.0)	8.0 (4.5)	8.5 (4.0)	0.572
Added Sugar (0–10)	10.0 (2.0)	9.0 (4.5)	10.0 (1.0)	0.173
Sodium (0–10)	9.0 (3.0)	8.0 (4.0)	10.0 (3.0)	0.059
Saturated Fat (0–10)	3.0 (4.0)	3.0 (5.5)	4.0 (4.0)	0.406
Fatty Acid (0–10)	0.0 (3.0)	1.0 (3.0)	0.0 (3.3)	0.146

Parametric variables are presented as mean ± SD and non-parametric variables as median (interquartile range).
Mean FEV_1%: 77.4 ± 25.1 %.
Of participants who completed the three-day food diary, $n = 1$ did not report their FEV_1%.
P-values were derived with independent sample *t*-tests (Non-parametric: Mann Whitney *U* test).
Significance was derived from a *p*-value of <0.05.
* Non-parametric test.
Abbreviations: FEV_1%, forced expiratory volume as a percentage predicted; HEI-2020, Healthy Eating Index 2020; SD, standard deviation.

4. Discussion

This study provides a comprehensive evaluation of dietary intakes of adult PwCF, presenting findings that are theoretically relevant for future nutrition practices, guidelines, and interventions. This study uniquely examines diet quality in a European context, where environmental influences on diet and food choice may differ to that of previous research assessing diet quality in CF from other regions [48–52]. The results indicate that the diets of Irish adults with CF are inadequate to meet their health needs and may place them at risk of adverse metabolic outcomes.

While the median percentage EAR of adult PwCF is within the range recommended by the European CF-specific nutrition guidelines [7], 50 % of participants have percentage EAR below this recommendation. A recent systematic review ($N = 19$ cohorts) evaluating the dietary intakes and diet quality of adult PwCF worldwide ($N = 724$) observed that of studies that reported percentage EAR ($n = 9$), based on mean and median intake data most cohorts were achieving the recommendation ($n = 9$). However, most studies in this review were from a pre-modulator era, with no study in the systematic review provided information on the percentage of participants in each cohort achieving the recommendation [28]. The low percentage of individuals in this study meeting the EAR target may be attributed to the use of modulator therapy, whereby the population requires less energy intake than before to maintain their BMI [7]. As a result, they may be consuming fewer calories overall, which is further highlighted by the fact that only one participant in this study is classified as underweight. However, the limited data available from previous research makes it difficult to confirm this hypothesis definitively. Furthermore, the current study also aligns with the findings presented in Greaney et al. [28] in relation to mean and median macronutrient and micronutrient intake distributions, with %TEI fat, saturated fat and sugar all above nutrition recommendations[39,41], fibre and fat soluble vitamin D, E and K_1 intakes suboptimal compared to DRVs [39] and %TEI protein sufficient alongside intakes of calcium, and zinc relative to age and gender specific reference intakes [39]. However, results differ in that the group mean %TEI carbohydrates is below guideline recommendations [39], while in Greaney et al. [28], 80 % of cohorts were achieving recommendations. In a representative sample of the Irish general population the Irish National Adult Nutrition Survey II reflected similar consumption patterns in relation to mean %TEI carbohydrates, protein, fat, total sugar and saturated fat [53].

High fat intakes are likely a result of ingrained and life-long dietary practices amongst PwCF, with long-standing clinical practice being to prescribe a diet high in energy and fat alongside pancreatic enzyme replacement therapy to achieve gender-specific BMI targets [8]. This has been suggested in other research assessing intakes amongst children living with CF where significantly higher EDNP food intakes were reported compared to controls, largely contributing to higher %TEI fat and energy intakes [54]. VSTs are now an established part of the management plan for many PwCF leading to significant changes in energy intake needs due to better lung function and nutrient absorption [55–58]. In light of the Academy of Nutrition and Dietetics recommendation that high fat diets are not therapeutically beneficial beyond meeting high energy targets [9], it may be timely to reconsider traditional dietary priorities for this population in line with the latest European CF-specific nutrition guidelines which advise that most PwCF follow similar macronutrient targets to general population nutrition guidelines [7]. However, higher energy intakes may still be required for specific patients (e.g., PI, low FEV_1%, BMI <20 kg/m^2, CF liver disease, etc.) highlighting that a more individualised approach to dietary advice is necessary in optimising the health and wellbeing of all PwCF.

Poor diet quality is reflected in food group consumption patterns, with a disproportionally high intake of EDNP foods and fats, spreads and oils recorded, alongside suboptimal intakes of vegetables, salad and fruit [42], aligning with inadequacies presented for micronutrient intakes (i.e., iron, vitamin D, E, K_1 and C). The reliance on EDNP foods amongst

PwCF has been replicated in previous research in both adult [50,51] and child CF populations [54]. With sustained intakes of EDNP foods closely associated with the development of diet-related chronic diseases in the general population [59], for many PwCF there is a necessity in rebalancing current intake patterns for the betterment of overall health and longevity. However, this pattern of eating is not exclusively present in PwCF, with various national dietary surveys displaying that similar dietary structures are present in the general population [53,60,61]. PwCF live in the same obesogenic environments [62] as the general population and experience the same marketing and commercial influences that encourage EDNP foods which can powerfully affect food choice [63–65]. Adherence to nutrition guidelines in the general population have sequentially proven difficult to achieve due to these intrinsic and extrinsic factors [66–68]. Where once, emphasis may not have been placed on this, in the CF modulator era, where BMI and metabolic syndromes are increasing [14,23], education and guidance is needed to promote healthier alternatives for PwCF. With the latest European CF nutrition guidelines advocating for more healthy dietary pattern consumption [7], the development of effective strategies for instilling dietary changes towards the achievement of a high-quality diet are warranted. Therefore, future research should aim to assess the enablers and barriers for PwCF to achieve the recommended food serves or dietary patterns promoted over the course of updated guideline implementation.

The mean HEI-2020 score achieved by participants falls within the lowest grade applicable (HEI-2020: 0–59) when compared to evaluation criteria [26], further indicating poor diet quality in Irish adult PwCF. This aligns with previous research in adult PwCF evaluating diet quality via the Healthy Eating Index – 2015 (HEI-2015) [48,49], which additionally reported significantly worse mean diet quality scores compared to the population level mean score (HEI-2015 score: 59, $p < 0.0001$)[60] and an age-matched control group (63.9 ± 27.3, $p < 0.001$) [49]. The most recent HEI-2020 scores reported for adults in the USA (19–30 years: 55.3; 31–59 years: 55.3; ≥60 years: 59.5) similarly are within the lowest HEI-2020 grade, but score notably lower in ages 19–59 years [27]. Furthermore, in a healthy population of adults in the USA ($n = 6614$, 30–74 years old), higher HEI-2015 diet quality scores were associated with significantly lower predicted 10-year cardiovascular disease risk and lower heart age, highlighting the importance of diet quality in lowering the risk of adverse cardiometabolic complications [69]. In the absence of evidence suggesting that higher-quality diets are ineffective for PwCF, similar dietary principles could be effectively applied within this population.

Examining the relationship between diet quality and lung function in adult PwCF, participants with a higher FEV_1% have significantly higher HEI-2020 diet quality scores, alongside significantly higher HEI-2020 total fruits and whole fruits component scores. Additionally, participants below the mean FEV_1% have a mean HEI-2020 score that falls within the lowest grade applicable to the index, with those above the mean FEV_1% achieving a mean HEI-2020 score above this classification. The results display that increases in FEV_1% appear to coincide with better diet quality, likely through increased intakes of fruits and fibre in individuals with higher FEV_1%. While the data indicate a significant difference between those above and below the mean FEV1%, the clinical implications of these findings remain unclear. Caution is warranted in interpreting these results, as it should not be assumed that all PwCF with a higher FEV_1% have better diet quality. Consequently, a more thorough investigation of the relationship between diet quality and FEV_1% is needed to draw appropriate conclusions.

The use of a diet quality score within CF contexts may be advantageous moving forward in creating and measuring effects of future nutrition interventions and practices on disease progression and metabolic risk factors and may enable a deeper understanding of diet in CF patients beyond nutrient and food group intake data. However, utilising the HEI-2020 requires significant effort and can be quite time-consuming, making it challenging to implement broadly in clinical environments with strict time limitations. In these settings, a more fitting strategy to assess diet quality may be to continue evaluating dietary intakes against established national and regional nutrition guidelines.

This study presents a sample size larger than any assessing dietary intakes of adult PwCF from one country, with only Moen et al. [70] ($n = 136$), surpassing the number of participants involved when including studies which assessed dietary intakes of PwCF across multiple nations. Although imbalances related to PI status and modulator use were present, proportions are considered clinically representative, and based on projections for BMI, FEV_1% and other relevant clinical endpoints in registry data, the cohort is representative of the overall well population of PwCF moving forward [14] as well as many CF cohorts in Europe, Australia and USA [14,71,72]. While a subset of participants were not on CFTR modulators, exploratory comparisons suggested that their inclusion did not skew the overall findings (Appendix 3). However, the sample size disparity between those on and not on modulators should be acknowledged, and future studies may benefit from stratified recruitment or subgroup analysis to examine dietary differences by modulator status more robustly. While it must be recognised that the sample size in this study is still not representative, it is important to acknowledge that a wide and strategic recruitment process was employed. For future studies, achieving a larger and more representative sample may require broader recruitment across countries with similar habitual dietary patterns (e.g., the UK and Northern Europe), continued use of both online and paper-based data collection methods, and benchmarking participant characteristics against CF registry data. Additionally, recognition of the time commitment required from participants in completing detailed dietary assessments may be important to enhance engagement and retention. Furthermore, proportions of females in this population are higher than the most recent Irish registry data report (43.6 %) [14] which may have impacted results based on typical differences observed in dietary intakes between males and females [53]. Data were collected using self-report measures which have limitations in terms of recall bias and the reporting of conditions like CF bone and liver disease may have been subject to misclassification bias due to variations in how these conditions were defined. Data were not collected on date of diagnosis, mutation type, and the specific VSTs used by each participant. Therefore, assessment of dietary intakes and quality against diagnosis stage and comparisons of dietary intakes and quality between mutations and type of VSTs were not possible. Lastly, contributions from micronutrient supplements were not included. However, the primary focus was to assess dietary intake sufficiency without the influence of supplements to provide a more explicit picture of diet quality and adherence to dietary guidelines specifically from food sources.

It is clear from this cross-sectional study that the diet quality of adult PwCF is suboptimal, with misalignments in macronutrient, micronutrient, and food group intakes relative to CF-specific, national, and European guidelines. While the cohort displayed clinical measurements representative of an overall well CF population, many did not meet CF-specific minimum energy requirements despite a significant proportion of food eaten coming from the EDNP food category, which contributed to excesses in %TEI fat, saturated fat, and sugar. With current dietary patterns in CF linked to diet-related chronic diseases in the general population and rising incidences amongst PwCF, emphasis on diet quality is of paramount importance. Future research should validate HEI-2020 in a representative CF sample size and explore barriers to dietary changes. Important factors to consider include clinical equipoise, food insecurity and the time and treatment burdens PwCF face. While changing ingrained dietary habits is complex, understanding these barriers will be essential for developing patient-informed nutrition guidelines and interventions tailored to the unique challenges of this population.

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Data availability

The data that support the findings of this study are available from the corresponding author upon reasonable request.

CRediT authorship contribution statement

Cian Greaney: Conceptualization, Methodology, Validation, Formal analysis, Investigation, Resources, Data curation, Writing – original draft, Writing – review & editing, Visualization, Supervision, Project administration, Funding acquisition. **Ellen McCarthy:** Validation, Formal analysis, Data curation. **Lauren O'Brien:** Validation, Formal analysis, Data curation. **Sarah Tecklenborg:** Resources, Writing – review & editing, Funding acquisition. **Ciara Howlett:** Resources, Writing – review & editing. **Karen Cronin:** Resources, Writing – review & editing. **Clodagh Landers:** Resources, Writing – review & editing. **Mary Connolly:** Resources, Writing – review & editing. **Derbhla O'Sullivan:** Resources, Writing – review & editing. **Aoife Whiston:** Formal analysis, Writing – review & editing. **Katie Robinson:** Conceptualization, Writing – review & editing, Supervision, Funding acquisition. **Audrey Tierney:** Conceptualization, Methodology, Resources, Writing – review & editing, Supervision, Project administration, Funding acquisition.

Declaration of competing interest

The authors declare the following financial interests/personal relationships which may be considered as potential competing interests: Sarah Tecklenborg is a staff member of Cystic Fibrosis Ireland, a contributing funder of the project, with Cystic Fibrosis Ireland representing the views of the Cystic Fibrosis community on the steering committee and facilitating elements of the study. No other authors declare any conflicts of interest.

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Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.jcf.2025.07.004.

References

[1] Grasemann H, Ratjen F. Cystic fibrosis. N Engl J Med 2023;389(18):1693–707.
[2] Bailey J, Rozga M, McDonald CM, Bowser EK, Farnham K, Mangus M, et al. Effect of CFTR modulators on anthropometric parameters in individuals with cystic fibrosis: an evidence analysis center systematic review. J Acad Nutr Diet 2021;121(7):1364–78.
[3] Coriati A, Ma X, Sykes J, Stanojevic S, Ruseckaite R, Lemonnier L, et al. Beyond borders: cystic fibrosis survival between Australia, Canada, France and New Zealand. Thorax 2023;78(3):242–8.
[4] Stephenson AL, Sykes J, Stanojevic S, Quon BS, Marshall BC, Petren K, et al. Survival comparison of patients with cystic fibrosis in Canada and the United States: a population-based cohort study. Ann Intern Med 2017;166(8):537–46.
[5] Kawchak DA, Zhao H, Scanlin TF, Tomezsko JL, Cnaan A, Stallings VA. Longitudinal, prospective analysis of dietary intake in children with cystic fibrosis. J Pediatr 1996;129(1):119–29.
[6] Saxby N, Painter C, Kench A, King S, Crowder T, van der Haak N, et al. Nutrition guidelines for cystic fibrosis in Australia and New Zealand. Sydney: Thoracic Society of Australia and New Zealand; 2017.
[7] Wilschanski M, Munck A, Carrion E, Cipolli M, Collins S, Colombo C, et al. ESPEN-ESPGHAN-ECFS guideline on nutrition care for cystic fibrosis. Clin Nutr 2024;43(2):413–45.
[8] Stallings VA, Stark LJ, Robinson KA, Feranchak AP, Quinton H. Clinical practice guidelines on growth and nutrition subcommittee, et al. evidence-based practice recommendations for nutrition-related management of children and adults with cystic fibrosis and pancreatic insufficiency: results of a systematic review. J Am Diet Assoc 2008;108(5):832–9.
[9] McDonald CM, Alvarez JA, Bailey J, Bowser EK, Farnham K, Mangus M, et al. Academy of nutrition and dietetics: 2020 cystic fibrosis evidence analysis center evidence-based nutrition practice guideline. J Acad Nutr Diet 2021;121(8):1591–636. e3.
[10] Turck D, Braegger CP, Colombo C, Declercq D, Morton A, Pancheva R, et al. ESPEN-ESPGHAN-ECFS guidelines on nutrition care for infants, children, and adults with cystic fibrosis. Clin Nutr 2016;35(3):557–77.
[11] Hanna RM, Weiner DJ. Overweight and obesity in patients with cystic fibrosis: a center-based analysis. Pediatr Pulmonol 2015;50(1):35–41.
[12] Panagopoulou P, Fotoulaki M, Nikolaou A. Nousia-Arvanitakis S. prevalence of malnutrition and obesity among cystic fibrosis patients. Pediatr Int 2014;56(1):89–94.
[13] The Cystic Fibrosis Registry of Ireland (CFRI). 2023 Annual report CF registry of Ireland. 2023.
[14] Cystic Fibrosis Foundation (CFF). Patient registry 2022 Annual data report Bethesda, Maryland; 2023.
[15] Duckers J, Lesher B, Thorat T, Lucas E, McGarry LJ, Chandarana K, et al. Real-world outcomes of ivacaftor treatment in people with cystic fibrosis: a systematic review. J Clin Med 2021;10(7).
[16] Borowitz D, Lubarsky B, Wilschanski M, Munck A, Gelfond D, Bodewes F, et al. Nutritional status improved in cystic fibrosis patients with the G551D mutation after treatment with ivacaftor. Dig Dis Sci 2016;61(1):198–207.
[17] King SJ, Tierney AC, Edgeworth D, Keating D, Williams E, Kotsimbos T, et al. Body composition and weight changes after ivacaftor treatment in adults with cystic fibrosis carrying the G551 D cystic fibrosis transmembrane conductance regulator mutation: a double-blind, placebo-controlled, randomized, crossover study with open-label extension. Nutrition 2021;85:111124.
[18] Wainwright CE, Elborn JS, Ramsey BW, Marigowda G, Huang X, Cipolli M, et al. Lumacaftor–ivacaftor in patients with cystic fibrosis homozygous for Phe508del CFTR. N Engl J Med 2015;373(3):220–31.
[19] Southern KW, Murphy J, Sinha IP, Nevitt SJ. Corrector therapies (with or without potentiators) for people with cystic fibrosis with class II CFTR gene variants (most commonly F508del). Cochrane Database Syst Rev 2020;(12).
[20] Middleton PG, Mall MA, Dřevínek P, Lands LC, McKone EF, Polineni D, et al. Elexacaftor–tezacaftor–ivacaftor for cystic fibrosis with a single Phe508del allele. N Engl J Med 2019;381(19):1809–19.
[21] Carnovale V, Iacotucci P, Terlizzi V, Colangelo C, Medio P, Ferrillo L, et al. Effectiveness and safety of elexacaftor/tezacaftor/ivacaftor in patients with cystic fibrosis and advanced lung disease with the Phe508del/minimal function genotype. Respir Med 2021;189:106646.
[22] Heijerman HGM, McKone EF, Downey DG, Van Braeckel E, Rowe SM, Tullis E, et al. Efficacy and safety of the elexacaftor plus tezacaftor plus ivacaftor combination regimen in people with cystic fibrosis homozygous for the F508del mutation: a double-blind, randomised, phase 3 trial. Lancet 2019;394(10212):1940–8.
[23] Frost F, Nazareth D, Fauchier L, Wat D, Shelley J, Austin P, et al. Prevalence, risk factors and outcomes of cardiac disease in cystic fibrosis: a multinational retrospective cohort study. Eur Respir J 2023;62(4).
[24] Fung TT, Chiuve SE, McCullough ML, Rexrode KM, Logroscino G, Hu FB. Adherence to a DASH-style diet and risk of coronary heart disease and stroke in women. Arch Intern Med 2008;168(7):713–20.
[25] Harrington J, Fitzgerald AP, Layte R, Lutomski J, Molcho M, Perry IJ. Sociodemographic, health and lifestyle predictors of poor diets. Public Health Nutr 2011;14(12):2166–75.
[26] Krebs-Smith SM, Pannucci TE, Subar AF, Kirkpatrick SI, Lerman JL, Tooze JA, et al. Update of the healthy eating index: HEI-2015. J Acad Nutr Diet 2018;118(9):1591–602.
[27] Shams-White MM, Pannucci TE, Lerman JL, Herrick KA, Zimmer M, Meyers Mathieu K, et al. Healthy eating index-2020: review and update process to reflect the dietary guidelines for Americans, 2020-2025. J Acad Nutr Diet 2023;123(9):1280–8.
[28] Greaney C, Doyle A, Drummond N, King S, Hollander-Kraaijeveld F, Robinson K, et al. What do people with cystic fibrosis eat? Diet quality, macronutrient and micronutrient intakes (compared to recommended guidelines) in adults with cystic fibrosis–a systematic review. J Cyst Fibros 2023;22(6):1036–47.
[29] Minihane AM, Vinoy S, Russell WR, Baka A, Roche HM, Tuohy KM, et al. Low-grade inflammation, diet composition and health: current research evidence and its translation. Br J Nutr 2015;114(7):999–1012.
[30] Fritsch J, Garces L, Quintero MA, Pignac-Kobinger J, Santander AM, Fernández I, et al. Low-fat, high-fiber diet reduces markers of inflammation and dysbiosis and improves quality of life in patients with ulcerative colitis. Clin Gastroenterol Hepatol 2021;19(6):1189–99.

[31] Fung TT, McCullough ML, Newby PK, Manson JE, Meigs JB, Rifai N, et al. Diet-quality scores and plasma concentrations of markers of inflammation and endothelial dysfunction2. Am J Clin Nutr 2005;82(1):163–73.
[32] Bonhoure A, Boudreau V, Litvin M, Colomba J, Bergeron C, Mailhot M, et al. Overweight, obesity and significant weight gain in adult patients with cystic fibrosis association with lung function and cardiometabolic risk factors. Clin Nutr 2020;39(9):2910–6.
[33] Bass R, Alvarez JA. Nutritional status in the era of highly effective CFTR modulators. Pediatr Pulmonol 2024;59:S6–16.
[34] Cuschieri S. The STROBE guidelines. Saudi J Anaesth 2019;13(Suppl 1):S31–Ss4.
[35] Biasizzo A, Seljak BK, Valenčič E, Pavlin M, Santo Zarnik M, Blažica B, et al. An open-source approach to solving the problem of accurate food-intake monitoring. IEEE Access 2021;9:162835–46.
[36] Greaney C, Bohan K, Tecklenborg S, Casserly B, Green J, Van de Ven P, et al. Diet quality in cystic fibrosis–associations with patient reported outcome measures and enablers and barriers to eating a healthy diet: a protocol paper for a mixed methods study. HRB Open Res 2022;5:33.
[37] Etikan I, Musa SA. Alkassim RS. Comparison of convenience sampling and purposive sampling. Am J Theor Appl Stat 2016;5(1):1–4.
[38] Contreras-Bolívar V, Olveira C, Porras N, Abuín-Fernández J, García-Olivares M, Sánchez-Torralvo FJ, et al. Oral nutritional supplements in adults with cystic fibrosis: effects on intake, levels of fat-soluble vitamins, and bone remodeling biomarkers. Nutrients 2021;13(2):669.
[39] European Food Safety Authority (EFSA). Dietary reference values for nutrients. Summary Report. 2017.
[40] World Health Organization. Guideline: sugars intake for adults and children: world health organization; 2015 [Available from: https://www.who.int/publications/i/item/9789241549028/.
[41] Irish Heart Foundation. Nutrition guidelines for heart health: with policy recommendations 2007 [updated 2007. Available from: http://hdl.handle.net/10147/45656.
[42] Healthy Ireland; Department of health. the food pyramid gov.ie: department of health, 2016 [updated 3rd November 2020. Available from: https://www.gov.ie/en/publication/70a2e4-the-food-pyramid/#.
[43] Millar SR, Navarro P, Harrington JM, Perry IJ, Phillips CM. Dietary quality determined by the healthy eating index-2015 and biomarkers of chronic low-grade inflammation: a cross-sectional analysis in middle-to-older aged adults. Nutrients 2021;13(1):222.
[44] Navarro P, Mehegan J, Murrin CM, Kelleher CC, Phillips CM. Adherence to the healthy eating index-2015 across generations is associated with birth outcomes and weight status at age 5 in the lifeways cross-generation cohort study. Nutrients 2019;11(4):928.
[45] Food Patterns Equivalents Database (FPED), [Internet]. 2021 [cited 03 October 2023]. Available from: www.ars.usda.gov/Services/docs.htm?docid=23871.
[46] National Cancer Institute. Healthy eating index. overview & background of the healthy eating index [cited 25 November 2024. Available from: https://epi.grants.cancer.gov/hei/.
[47] Weir CB, Jan A. BMI classification percentile and cut off points. Jun 26. StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023. 2025 Jan-Available from, https://www.ncbi.nlm.nih.gov/books/NBK541070/.
[48] Bass RM, Tindall A, Sheikh S. Utilization of the healthy eating index in cystic fibrosis. Nutrients 2022;14(4):834.
[49] Bellissimo MP, Zhang I, Ivie EA, Tran PH, Tangpricha V, Hunt WR, et al. Visceral adipose tissue is associated with poor diet quality and higher fasting glucose in adults with cystic fibrosis. J Cyst Fibros 2019;18(3):430–5.
[50] Armaghanian N, Atkinson F, Taylor N, Kench A, Brand-Miller J, Markovic T, et al. Dietary intake in cystic fibrosis and its role in glucose metabolism. Clin Nutr 2020;39(8):2495–500.
[51] Tierney AC, Fong-To SA, Clode M, Casamento J, Tsee Woon Yuen A, King S. Vegetable, fruit, dairy and discretionary food intake of cystic fibrosis (CF) patients and comparison with the general population: a cross-sectional study. J Cyst Fibros 2015;14:S118.
[52] King SJ, Thompson S, Taylor G, Malesic L, George ES, Tierney AC. The feasibility and applicability of the healthy eating index for Australian Adults (HEIFA-2013) score for dietary assessment in adults with cystic fibrosis. J Cyst Fibros 2018;17:S111.
[53] Irish Universities Nutrition Alliance (IUNA). National adult nutrition survey II: main report. 2024.
[54] Sutherland R, Katz T, Liu V, Quintano J, Brunner R, Tong CW, et al. Dietary intake of energy-dense, nutrient-poor and nutrient-dense food sources in children with cystic fibrosis. J Cyst Fibros 2018;17(6):804–10.
[55] Matel JL, Milla CE. Nutrition in cystic fibrosis. Seminars in respiratory and critical care medicine, 2009. © Thieme Medical Publishers; 2009.
[56] Zemanick ET, Harris JK, Wagner BD, Robertson CE, Sagel SD, Stevens MJ, et al. Inflammation and airway microbiota during cystic fibrosis pulmonary exacerbations. PloS One 2013;8(4):e62917.
[57] Rowe SM, Heltshe SL, Gonska T, Donaldson SH, Borowitz D, Gelfond D, et al. Clinical mechanism of the cystic fibrosis transmembrane conductance regulator potentiator ivacaftor in G551D-mediated cystic fibrosis. Am J Respir Crit Care Med 2014;190(2):175–84.
[58] Gelfond D, Ma C, Semler J, Borowitz D. Intestinal pH and gastrointestinal transit profiles in cystic fibrosis patients measured by wireless motility capsule. Dig Dis Sci 2013;58(8):2275–81.
[59] Joint WHO/FAO Expert Consultation (WHO and FAO). Diet, nutrition and the prevention of chronic diseases. Tech Rep Ser - World Health Organ Geneva 2003;916(i–viii):1–149.
[60] U.S. Department of Agriculture (USDA). U.S. department of health and human services (HHS). Dietary guidelines for Americans, 2020-2025. 2020.
[61] Bates B, Collins D, Jones K, Page P, Roberts C, Steer T, et al. NDNS: results from years 9 to 11 (2016 to 2017 and 2018 to 2019). Public Health Engl 2020.
[62] Swinburn BA, Kraak VI, Allender S, Atkins VJ, Baker PI, Bogard JR, et al. The global syndemic of obesity, undernutrition, and climate change: the Lancet commission report. Lancet 2019;393(10173):791–846.
[63] Harris JL, Fleming-Milici F. Food marketing to adolescents and young adults: skeptical but still under the influence. The psychology of food marketing and overeating. Routledge; 2019. p. 25–43.
[64] Larson NI, Story MT, Nelson MC. Neighborhood environments: disparities in access to healthy foods in the US. Am J Prev Med 2009;36(1):74–81.
[65] Harris JL, Bargh JA, Brownell KD. Priming effects of television food advertising on eating behavior. Health Psychol 2009;28(4):404.
[66] Banfield EC, Liu Y, Davis JS, Chang S, Frazier-Wood AC. Poor adherence to US dietary guidelines for children and adolescents in the national health and nutrition examination survey population. J Acad Nutr Diet 2016;116(1):21–7.
[67] Briggs R, Canney M, Carey D, Donoghue O, Foley M, Kenny RA, et al. Health and wellbeing: active ageing for older adults in Ireland. Trinity College Dublin: The Irish Longitudunal Study on Aging; 2017. Report No.: 978-1-907894-13-8.
[68] Healthy Ireland Survey 2021: Summary Report. Ipsos mrbi. Department of Health; 2021.
[69] Zhang Y, Lu C, Li X, Fan Y, Li J, Liu Y, et al. Healthy eating index-2015 and predicted 10-year cardiovascular disease risk, as well as heart age. Front Nutr 2022;9:888966.
[70] Moen IE, Nilsson K, Andersson A, Fagerland MW, Fluge G, Hollsing A, et al. Dietary intake and nutritional status in a Scandinavian adult cystic fibrosis-population compared with recommendations. Food Nutr Res 2011;55.
[71] Zolin A., Orenti A., Jung A., van Rens J., et al. ECFSPR annual report 2021. 2023.
[72] Ahern S, Pourghaderi AR, Ruseckaite R, Caruso M, Liman J, Wark P, et al. The ACFDR registry annual report, 2023. Monash University: School of Public Health and Preventive Medicine; 2024. Report No.: 25.