CLINICAL NUTRITION
DOI: 10.12957/demetra.2025.74356

DEMETRA
Alimentação, Nutrição & Saúde

e-ISSN:2238-913X

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Article from the Projeto de Iniciação Cientifica (PIBIC, Scientific Initiation Project) - Influence of nutritional supplementation on the quality of life in children and adolescents with cystic fibrosis treated at a Reference Hospital in Southern Brazil, by the author Mônica Ribeiro de Moraes, from the University of Southern Santa Catarina, from 2019 to 2020.

Funding: This study was funded by FAPESC Public Call No. 03/2017, process number 23038.013359/2017-71 and developed within the scope of the Graduate Program in Health Sciences of the Universidade do Sul de Santa Catarina, Tubarão-SC, Brazil.

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Influence of the use of hypercaloric oral nutritional supplement on the quality of life in children and adolescents with cystic fibrosis

Influência do uso de suplemento nutricional oral hipercalórico na qualidade de vida em crianças e adolescentes com fibrose cística

Abstract

Introduction: Cystic fibrosis (CF) is a chronic, genetic, and progressive disease, and early diagnosis and treatment developments have increased life expectancy. *Objective:* To evaluate the influence of high-calorie oral nutritional supplements on the quality of life (QoL) of children and adolescents with CF treated at a referral hospital in southern Brazil. *Methods:* This was a cross-sectional study of CF patients aged 2 to 14 years. Sociodemographic data, clinical characteristics, high-caloric oral nutritional supplement use, and QoL were reviewed using the Cystic Fibrosis Questionnaire (versions for children aged 6 to 11 years, 12 to 13 years, and ≥14 years) and the Pediatric Quality of Life Inventory (versions for children aged 2 to 4 years and 5 to 7 years). Study participants were categorized according to the age range of the relevant questionnaires. *Results:* Eighty-two patients participated in the study, of whom 73.2% used nutritional supplements. When comparing nutritional supplement use with QoL, children aged 2 to 4 years who used supplements had higher scores in the emotional domain. Among those aged 6 to 11 years, those who did not use nutritional supplements had higher scores in the physical domains: "body image," "food," "treatment," and "weight." Among 14-year-old adolescents, those using nutritional supplements exhibited lower scores in the "treatment" domain and higher scores in the "vitality" domain. *Conclusion:* The results of this study indicate that nutritional supplement use can influence different dimensions of QoL in different age groups.

Keywords: Cystic fibrosis. Nutritional supplements. Quality of life.

Resumo

Introdução: A fibrose cística (FC) é uma doença crônica, genética e progressiva, e seu diagnóstico precoce e a evolução do tratamento têm aumentado a expectativa de vida dos pacientes. *Objetivo*: Avaliar a influência do uso de suplemento nutricional oral hipercalórico na qualidade de vida (QV) em crianças e adolescentes com FC atendidos em um Hospital de Referência do Sul do Brasil. *Métodos*: Estudo transversal que analisou pacientes com idade entre 2 e 14 anos com FC. Foram analisados dados sociodemográficos,

características clínicas, uso de suplemento nutricional oral hipercalórico e QV por meio dos questionários *Cystic Fibrosis Questionnaire*, nas versões de 6 a 11 (versão para pais), 12 a 13 anos (versão para adolescentes) e ≥ 14 anos (versão para adolescentes); e *Pediatric Quality of Life Inventory*, nas versões de 2 a 4 anos e 5 a 7 anos (versão para pais). Os participantes do estudo foram categorizados segundo a faixa etária dos respectivos questionários. ***Resultados***: Participaram do estudo 82 pacientes, dos quais 73,2% utilizam suplemento nutricional. Ao compararmos o uso de suplemento nutricional com a QV, as crianças de 2 a 4 anos que utilizam suplemento tiveram maior pontuação no domínio emocional. Naqueles entre 6 a 11 anos, os que não utilizavam suplemento nutricional apresentaram maiores pontuações nos domínios físico, "imagem corporal", "alimentação", "tratamento" e "peso". Nos adolescentes de 14 anos, aqueles que fazem uso de suplemento nutricional apresentaram menor pontuação no domínio "tratamento" e maior pontuação no domínio "vitalidade". ***Conclusão***: Os resultados deste estudo indicam que o uso de suplemento nutricional pode influenciar diferentes dimensões da QV em distintas faixas etárias.

Palavras-chave: Fibrose cística. Suplementos nutricionais. Qualidade de vida.

INTRODUCTION

Cystic fibrosis (CF) is an inherited, autosomal recessive and potentially fatal disease characterized by chronic lung disease, pancreatic insufficiency, and malnutrition. It is caused by mutations in the gene located on the long arm of chromosome 7, which transcribes the Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) protein.[1,2] These mutations lead to a deficiency or absence of functional CFTR proteins in the apical membrane of epithelial cells in multiple body systems, resulting in the accumulation of thick, sticky mucus in the lungs and pancreas.[2]

The incidence of cystic fibrosis varies among different ethnic groups, being more prevalent in individuals of Caucasian descent. According to data from the Cystic Fibrosis Foundation (2023), it is estimated that approximately 70,000 people are affected by the disease worldwide. In Brazil, the prevalence is approximately 1 case per 10,000 live births; this rate varies regionally though,being higher in the southern states (1 in 2,400 live births).[3]

The concept of quality of life is widely used when referring to the management of chronic diseases. However, it is a subjective data, which must be evaluated technically and specifically, to describe a way to ensure treatment that increases survival and provides a better quality of life (QoL) in the physical, psychological, and social aspects.[4] The literature shows that CF patients who experience more pulmonary exacerbations and have a lower body mass index (BMI) have a worse QoL.[5,6]

CF individuals have high energy needs due to their high caloric expenditure but many patients fail to meet their daily calorie intake. In this connection it is essential to consider complementary strategies to optimize nutritional intake, such as the use of oral nutritional supplements (ONS).[7,8]

Growing evidence suggests that ONS, by providing macronutrients and micronutrients for special medical purposes in addition to the usual diet, can contribute to improving the clinical conditions of malnourished in-patients. Studies indicate that prolonged use of ONS in CF patients, aimed at increasing the energy density and protein content of their diet improves their nutritional conditions and, consequently, their quality of life.[7,8]

The aim of this study was to evaluate the influence of hypercaloric oral nutritional supplements on the QoL of CF children and adolescents treated at a referral hospital in southern Brazil.

METHODS

This is a cross-sectional study with convenience sampling conducted at the CF outpatient clinic of a public hospital in southern Brazil from May 2019 to March 2020. As a referral center for cystic fibrosis patients, the clinic has a specialized multidisciplinary team, ensuring comprehensive, high-quality patients' care. Services offered include pulmonology, gastroenterology, nutrition, endocrinology, physical therapy, nursing, psychology, and social work, providing comprehensive patients' care.

Patients receive regular visits every three months, during which they undergo evaluations across various specialties. Regarding nutritional support, all study participants are monitored by a nutritionist, who, after an individualized assessment, determines the need for and prescribes ONS, aiming to optimize the nutritional status and quality of life of those patients.

The population consisted of children and adolescents of both genders, aged between 2 and 14 years, having a confirmed CF diagnosis. The sample for this study was defined using the non-probabilistic convenience method, such as a census.

The sample consisted of 117 patients treated at the outpatient clinic. After applying the inclusion and exclusion criteria, 90 patients remained, of which eight refused to participate. Thus, 82 participants comprised the final sample.

Children and adolescents whose parents and/or guardians refused to answer the questionnaires or did not allow any of the planned anthropometric measurements were excluded. Children under 2 years of age and adolescents in the hospital transition phase who had recently turned 15 were also excluded from the sample.

Parents or guardians signed a written informed consent form prior to the study, and children 6 years of age and older and adolescents signed an assent form, agreeing to participate in the study.

The study was approved by the Research Ethics Committee of the university proposing the study (CAAE00189418.6.0000.5369), and of the hospital responsible for the outpatient clinic (CAAE 00189418.6.3001.5361).

A questionnaire developed by the study authors was applied; it contained sociodemographic items – age (years), gender (male and female), ethnicity (white, black and mixed race), age at diagnosis (months), type of CFTR mutation (F508del homozygous, F508del heterozygous, other mutations), and use of hypercaloric oral nutritional supplement (yes and no).

Patients who used the product Fortini Plus MultiFiber® (liquid version) a ready-to-use supplement that has a completeset of nutrients and is high in calories (1.5 kcal/ml), were considered to be on ONS. Intended for children aged 3 to 10 years this supplement has a balanced composition of 10% protein, 50% carbohydrates, 40% fat, and is gluten- and lactose-free. Patients who used Frebini Energy Drink® (liquid version), a ready-to-use supplement containing a set of complete nutrients too and is high in calories (1.5 kcal/ml), were considered to be on ONS. Intended for children aged 1 to 12 years, it has a balanced composition of 10.2% protein, 49.8% carbohydrates, and 40% fat, and is also free of fiber and gluten.

Quality of life was assessed using the Cystic Fibrosis Questionnaire (CFQ), and the Pediatric Quality of Life Inventory (PedsQL), both previously validated in Brazil.[8,9]

The CFQ-R has four versions, three of which were used in this study: a version for parents of children 6 to 11 years old, administered to 32 children; a version for adolescents (12 to 13 years old), administered to 13 participants and a version for adolescents (≥ 14 years old), administered to five 14-year-old adolescents. The CFQ-R considers the physical, image, digestive, respiratory, emotional, social, eating, treatment, vitality, health, social role, and weight domains. Each domain has a score, and their sum generates the total score, whose values can range from 0 to 100 (where 100 corresponds to the most positive possible health-related quality of life [HRQoL]).[8]

To assess the HRQoL of children aged 2 to 5 years, since there is no specific QoL questionnaire for CF, the PedsQL was used. It has four versions. Two of them were used in this study: a version for children aged 2 to 4 years, administered to 28 participants, and for parents of children aged 5 to 7 years, administered to three 5-year-old participants. The PedsQL considers the physical, emotional, social, and academic domains. Each domain has a score, and their sum generates the total score, whose values can range from 0 to 100 (where 100 corresponded to the most positive possible HRQoL).[9]

Age groups were categorized in order to avoid an overlap between the age groups assessed by each questionnaire, ensuring greater accuracy in data analysis.

The data obtained were entered into an Excel program and exported to SPSS 18.0, where they were examined. The results were summarized as absolute numbers and percentages for nominal variables, mean and standard deviation, and minimum and maximum values for numerical variables. The Shapiro-Wilk normality test was performed. Bivariate analysis was performed using Pearson's chi-square test for qualitative variables and the Student's t-test or Mann-Whitney test for quantitative variables, as appropriate. Values of $p < 0.05$ were considered significant.

RESULTAS

Eighty-two children and adolescents with a confirmed diagnosis of CF were evaluated; 54.9% were male; 87.8% were white; their mean age was 7.37 ± 4.20 years. The mean age at diagnosis was 5.95 ± 17.11 months; the most prevalent mutation was homozygous F508del, with 44.4%, and 73.2% used ONS. When the groups were compared, a significant difference was observed according to gender and diagnosis, since in the group that did not use ONS, males predominated (77.3%, $p = 0.012$) and the highest mean age at diagnosis was 10.45 ± 23.19 months ($p = 0.015$). In the other variables, no statistically significant differences were observed (Table 1).

Table 1. Sociodemographic and clinical characteristics, according to the use of hypercaloric oral nutritional supplements in CF children and adolescents treated at a referral hospital in southern Brazil. Florianópolis-SC, 2019–2020.

Variable	Nutritional supplement		p-value
	Yes n (%)	Non (%)	
Gender			
Male	28 (46.7)	17 (77.3)	0.012
Female	32 (53.3)	5 (22.7)	
Ethnicity			
White	53 (88.3)	19 (86.4)	
Black	5 (8.3)	2 (9.1)	0.960
Brown	2 (3.3)	1 (4.5)	
Age*#	7.27±4.13	7.64±4.47	0.850
Age at diagnosis **#	4.30±14.15	10.45±23.19	0.015
Mutation			
Delta f508 homozigotous	28 (46.7)	8 (38.1)	0.591
Delta f508 heterozigotous	21 (35)	10 (47.6)	
Other mutations	11(18.3)	3 (14.3)	

Legend: *years; **months. # Results expressed as Mean ± Standard Deviation.
Source: Research data, prepared by the authors (2025).

In the assessment of the QoL of children aged 2 to 4 years in relation to the use of ONS, we observed a statistical difference only in the emotional domain (75 [65]; $p = 0.017$), since those using ONS obtained the highest score. All 5-year-old children used nutritional supplements – therefore, only the domains of the questionnaire were described (Table 2).

Table 2. Scores in the quality of life domains of the Pediatric Quality of Life Inventory according to the use of nutritional supplements by CF children treated at a referral hospital in southern Brazil. Florianópolis-SC, 2019–2020.

	PedsQL 2 to 4 years			PedsQL 5 years
Domains	Nutritional supplement		p-value	Nutritional supplement
	Median [IIQ]			Median [IIQ]
	Yes (n=19)	No (n=9)		Yes (n=3)
Physical	90.62 [81.25]	93.75 [79.68]	0.941	96.62 [81.25]
Emotional	75 [65]	70 [70]	0.017	75 [65]
Social	90 [85]	90 [77.5]	0.526	90 [85]
School	100 [91.66]	100 [70.83]	0.661	100 [91.66]

Legend: IQR = interquartile range (Q75-Q25)
Source: Research data, prepared by the authors (2025).

In the evaluation of the QoL of children and adolescents in relation to the use of ONS, a statistical difference was observed in the physical domains (91.62 [41.67]; p=0.006), body image (88.89 [100]; p=0.046), food (66.67 [100]; p=0.007), treatment (66.67 [77.78]; p=0.010) and weight (66.67 [100]; p=0.003), since those who used ONS obtained the lowest score (Table 3).

Table 3. Scores in the quality of life domains of the Cystic Fibrosis Questionnaire – version for parents of children aged 6 to 11, according to the rate of CF children and adolescents treated at a referral hospital in southern Brazil. Florianópolis-SC, 2019–2020.

Domains	Nutritional Supplement		p-value
	Yes	No	
	(n = 25)	(n = 7)	
	Median [IQR]		
Physical	91.67 [41.67]	100 [12.50]	0.006
Emotional	86.67 [46.67]	80 [26.67]	0.904
Vitality	80 [60]	86.67 [53.33]	0.304
Education	91.67 [75]	83.33 [33.33]	0.298
Body Image	88.89 [100]	100 [11.11]	0.046
Nutrition	66.67 [100]	83.33 [33.33]	0.007
Treatment	66.67 [77.78]	100 [55.56]	0.010
Health	77.78 [77.78]	88.90 [33.33]	0.124
Respiratory	88.89 [50]	83.33 [27.78]	0.736
Digestion	77.78[44.44]	77.78 [33.33]	0.750
Weight	67.67 [100]	100 [66.67]	0.003

Legend: IQR = interquartile range (Q75-Q25)
Source: Research data, prepared by the authors (2025).

In the assessment of the QoL of adolescents aged 12-13 years in relation to the use of nutritional supplements, no statistical difference was observed (p=0.05) (Table 4).

Table 4. Scores in the quality of life domains of the Cystic Fibrosis Questionnaire - 12–13 years version forCF adolescents treated at a referral hospital in Southern Brazil. Florianópolis-SC, 2019–2020.

Domains	Nutritional Supplement		p-value
	Yes (n = 8)	No (n = 5)	
	Median [IQR]		
Physical	80[100]	72.22 [44.45]	0.833
Emotional	79.17 [33.34]	79.17 [37.50]	0.935
Body Image	64.28 [88.89]	77.78 [66.67]	0.342
Nutrition	100 [55.56]	66.67 [88.89]	0.082
Treatment	83.33 [22.22]	77.78 [33.33]	0.434
Respiratory	83.33 [100]	75 [25]	0.503
Digestion	91.66 [100]	100 [33.33]	0.746
Social	73.81 [76.19]	61.90 [31.10]	0.339

Legend: IQR = interquartile range (Q75-Q25)
Source: Research data, prepared by the authors (2025).

In the assessment of the QoL of 14-year-old adolescents in relation to the use of ONS, a statistical difference was observed in the "vitality" domain (86.66 [20]; p=0.034). Adolescents who used ONS exhibited a higher score in this domain, but in the "treatment" domain (66.67 [22.22]; p=0.046), they exhibited a lower score (Table 5).

Table 5. Scores in the quality of life domains of the Cystic Fibrosis Questionnaire – 14-year old version forCF adolescents treated at a referral hospital in southern Brazil. Florianópolis-SC, 2019–2020.

Domains	Nutritional Supplement		p-value
	Yes (n = 4)	No (n = 1)	
	Median [IQR]		
Physical	77.08 [41.67]	100 [0]	0.277
Emotional	86.66 [20]	73.33 [0]	0.264
Vitality	83.33 [25]	41.67 [0]	0.034
Education	100 [50]	83.33 [0]	0.429
Body Image	83.33 [100]	100 [0]	0.277
Nutrition	83.33 [77.78]	100 [0]	0.497
Treatment	66.67 [22.22]	100 [0]	0.046
Health	38.88 [44.45]	77.78 [0]	0.157
Respiratory	86.11 [66.67]	66.67 [0]	0.468
Digestion	94.44 [22.22]	88.89 [0]	0.709
Weight	83.33 [100]	100 [0]	0.429
Social	61.11 [38.89]	72.22 [0]	0.480

Legend: IQR = interquartile range (Q75-Q25)
Source: Research data, prepared by the authors (2025).

The results obtained demonstrated that CF patients monitored at the reference center had a good QOL perception, which could be verified, in most of the domains assessed, as the scores were above 50 (Tables 3, 4 and 5).

DISCUSSION

In our study, a higher prevalence of male children and adolescents was observed, a finding that corroborates other Brazilian studies, as well as data from European countries.[3,10-12] Furthermore, the predominance of Caucasian ethnicity observed in this study is in agreement with data from the Brazilian CF Registry[3] and from the American Cystic Fibrosis Registry.[13]

According to the Brazilian CF Registry[3] most of the cystic fibrosis population is in the age group of 5 to 10 years, followed by the population under 5 years of age according to the data obtained in our study which also showed that the average CF diagnosis occurred at 6 months of age. According to the literature, the earlier the diagnosis, the better the prognosis and the longer the survival of CF individuals.[11,14]

The F508del mutation was detected in 44.4% of the children and adolescents in our study. This is the most common CF mutation, and according to the latest Brazilian CF registry,[3] it is present in 43.7% of CF individuals. This variant is associated with more severe clinical manifestations of the disease and poorer nutritional status.[3] In a study conducted in Campinas, São Paulo, the F508del mutation was present in 39.82% of cystic fibrosis patients.[15] In contrast, in Porto Alegre, Rio Grande do Sul, the mutation was present in 71.43% of cystic fibrosis patients; such difference may be associated with the ethnic difference of each region.[16]

In our study the prevalence of children and adolescents using ONS is high, corroborating other studies.[12,17] The literature demonstrates that nutrition is of great importance in the well-being and survival of cystic fibrosis patients, and it is widely established that malnutrition has an impact on the quality of life and prognosis of CF individuals.[17] As a consequence of the disease, cystic fibrosis patients experience an increased energy expenditure due to inflammation and lung infection, which leads to malnutrition.[11]

Many families of CF children and adolescents face difficulties in providing adequate nutritional support and maintaining good nutritional status in their offspring.[18] Furthermore, a major challenge for families is the inadequate eating behaviors, characteristic of this phase of childhood; hence the recommendation, in some cases, to use nutritional supplements, aiming to ensure patients the best clinical condition.[19]

QoL in this study was satisfactory regardless of the age group assessed. This may be explained by the fact that children are less anxious and depressed, and have greater optimism about coping with the disease, when compared to adults.[20] Symptoms of anxiety and depression are more frequently reported in adolescents and adults and are associated with lower scores in QoL reports.[20]

The higher scores in the physical, body image, nutrition, treatment, and weight domains among participants aged 6 to 11 who did not use nutritional supplements suggest that these individuals have better nutritional status and less clinical impairment. Children who do not require supplementation may have a dietary intake adequate for their energy needs, which promotes growth and development and has a positive impact on QoL. Furthermore, the progression of cystic fibrosis is heterogeneous, and individuals with a more stable clinical condition may maintain good health without requiring further nutritional interventions, which may positively impact their perception of QoL.[21]

Additionally, the perception of parents, who were in charge of completing the questionnaires in this age group, may influence the results. Parents of children who do not require supplementation may perceive their children as healthier and less impacted by the disease, which is reflected in more positive assessments in the

domains analyzed. Thus, the absence of the nutritional supplementation requirement may be an indirect indicator of better clinical and nutritional status, less emotional and social impact, and a lower perceived therapeutic burden, explaining the better scores in the physical, "body image," "nutrition," "treatment," and "weight" domains among children aged 6 to 11 who do not use nutritional supplementation.[22]

Fourteen-year-olds who used nutritional supplements scored higher in the "vitality" domain, while those who did not use ONS scored higher in the "treatment" domain. At this age, patients are beginning to take responsibility, at least in part, for their own treatment, which leads to greater knowledge about their disease. However, the rigid treatment routine ends up influencing daily activities and social relationships.[21]

In contrast, among adolescents aged 12 to 13, the use of ONS did not influence QoL, which may be related to several physiological, psychological, and social factors specific to this stage of the individual development. At this age, adolescents experience intense hormonal and emotional changes, which can affect their overall perception of health and well-being. Another relevant factor is that, in this age group, growth still occurs, but without the acceleration typical of late puberty. Thus, nutritional needs may not be as discrepant between those who use supplements and those who do not, minimizing the impact of these products on perceived QoL.[23,24]

Limitations of this study include the inability to perform a longitudinal assessment, as it is a cross-sectional study. There was no assessment of adherence to nutritional supplement use, housing conditions, access to food, basic sanitation, type of bacterial colonization, or pulmonary exacerbations. Furthermore, the small sample size is noteworthy, which is inherent to the clinical condition studied. This is a rare disease, and this study included all patients treated at this CF clinic, the only pediatric referral center in the Rio Grande do Sul state for the treatment of this disease. Another limitation of the study is the small number of adolescent participants, given that the time limit for treatment at the aforementioned hospital is up to 14 years of age.

Another important factor to consider is that the PedsQL is not a specific CF questionnaire, although it is a validated questionnaire that allowed the QoL assessment of CF children who are under 5 years of age, a fact little addressed in other studies, which helps making the results of our study more comprehensive.

CONCLUSION

The high prevalence of nutritional supplement use among children and adolescents with cystic fibrosis reflects the importance of nutritional support in that disease management. The findings of our study suggest that the relationship between supplement use and quality of life varies by age group, highlighting the complexity of the factors that influence the well-being of these patients. It is noteworthy that, in the case of children up to 11 years of age, the questionnaires were completed by their parents, which may have influenced the scores obtained, since the parents' perception of their children quality of life may differ from the children's actual perception. On the other hand, the adolescents responded directly to the questionnaire, which may explain the smaller difference between the groups and the possible influence of individual and subjective factors in assessing their own quality of life.

These results highlight the need for individualized strategies that take into account not only nutrition but also the emotional and psychosocial aspects of the disease treatment. Multidisciplinary monitoring, combined with an understanding of the different impacts of nutritional supplements on the quality of life for each age group, can contribute to more effective interventions, promoting greater treatment adherence and, consequently, a more positive prognosis for cystic fibrosis patients.

AKNOWLEDGMENTS

The authors gratefully acknowledge the Institutional Initiation Scholarship Program for the scientific initiation scholarship awarded to Monica Ribeiro de Moraes. Furthermore, the authors are grateful to the Santa Catarina State Research and Innovation Support Foundation for awarding a doctoral scholarship to Bruna Becker da Silva.

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Contributors

da Silva BB, de Moraes MR, Schlindwein AD, and Iser BPM participated in all stages, from the conception of the study to the review of the final version of the article.

Conflito de Interesses: The authors declare no conflict of interest.

Received: March 21, 2023
Accepted: July 1, 2025