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Nutritional Status, Muscle Strength and Functional Capacity in Children and Adolescents with Cystic Fibrosis

Abstract

Objective: To describe the prevalence of malnutrition in children and adolescents with cystic fibrosis (CF) at a reference center and to investigate the association between nutritional status, muscular strength and functional capacity in these individuals.

Method: Cross-sectional, observational and descriptive study. Malnutrition was defined according to body mass index (BMI) for age, height for age and arm muscle circumference (AMC) percentile. Manovacuometry, dynamometry and the 6-minute walk test (6MWT) were used to evaluate the respiratory and hand grip strength and functional capacity, respectively.

Result: A total of 57 patients, aged 13.26±3.1 years, 42.1% of the male gender were evaluated. When evaluating BMI/age, 59.6% of the patients presented nutritional risk and 22.8% were malnourished. Regarding height/age, 15.8% presented short stature for age and according to AMC, 66.7% of the sample showed to be nourished. In the analysis of the association of nutritional status with muscular strength and functional capacity, only a significant difference was observed between nourished and malnourished when nutritional status was evaluated by BMI/age. When comparing the distance walked in 6MWT, hand grip strength, maximum inspiratory pressure and maximum expiratory pressure, according to the categories of the BMI/age percentile, the malnourished walked less distance in 6MWT as well as presented lower values of muscular strength.

Conclusion: The impairment of nutritional status when assessed by the BMI/age index was associated with decreased functional capacity and upper limb muscle strength in children and adolescents with CF.

Keywords: Cystic fibrosis; Muscle strength; Nutritional status

Research Article

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Nelbe Nesi Santana*, Christine Pereira Gonçalves and Célia Regina Miranda de Moutinho Chaves

National Institute of Woman, Child and Adolescent Fernandes Figueira, Brazil

***Corresponding author:** Nelbe Nesi SantanaNational Institute of Woman, Child and Adolescent Fernandes Figueira, Brazil, Tel: (21)2554-1768; Email: nelbenesi@iff.fiocruz.br

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Introduction

Cystic fibrosis (CF) is an autosomal recessive genetic disorder, characterized by dysfunction of Cystic Fibrosis Transmenbrane Conductance Regulator (CFTR), responsible for regulating the transport of sodium, chlorine and water through the membranes of epithelial cells [1,2]. The clinical manifestation of CF is multisystemic and the classic triad consists of chronic lung disease, pancreatic insufficiency and high concentrations of chloride in sweat [3]. Among the affected systems are the respiratory, digestive and musculoskeletal systems [4,5].

Nutritional status is one of the factors that may influence the prognosis of CF. It is one of the predictors of survival and is directly associated with pulmonary function, and consequently the morbidity and mortality of these patients [6,7]. Patients with CF may present impaired nutritional status with consequent reduction in muscle mass and strength, as well as a decline in lung function, which may contribute to fatigue both during exercise and during daily activities [8].

The aim of the study was to describe the prevalence of malnutrition in children and adolescents with CF attended at a reference center for the disease and to investigate the association between nutritional status, muscle strength and functional capacity in these Individuals. The hypothesis It is that children with good nutritional status are better able to perform activities of their daily life and respiratory and hand grip strength (HGS) than those that are considered malnourished.

Materials and Methods

A prospective, observational and descriptive study was carried out at the National Institute of Health of Women, Children and Adolescent Fernandes Figueira, which is a reference center for the pediatric treatment of CF patients In State of Rio de Janeiro and currently has 176 patients with this disease.

The data was collected from March to October 2016, on the day of the follow-up consultation at the Institute. Children and adolescents between 8 and 19 of age were included, with a diagnosis of CF confirmed by sweat test (≥ 60 mmol/L chlorine) in two samples and/or presence of two mutations in the CFTR gene, according to the consensus of Cystic Fibrosis Foundantion [9]. We excluded from the study those subjects with acute phase disease,

chronic hypoxemia with oxygen dependence, or with some condition that made the procedures impossible. The study was approved by the Ethics Committee in Research with human beings of the institution, by the number CAEE 52272115.0.0000.5269.

The weight (kg) was measured without shoes and with the minimum of clothes in a anthropometric platform scale (Filizola, São Paulo, Brazil) and the height (cm) in a stadiometer coupled to the scale, with the head position adjusted to the Frankfurt plan. The body mass index (BMI) was calculated by the weight divided by the height squared (kg/m^2). Arm circumference (AC) was measured at the midpoint of the right arm, relaxed, between the acromial point of the scapula and the olecranon of the ulna. The triceps skinfold (TS) was measured with a Lange Adipometer ® (Cambridge Scientific Industries, Cambridge, MD, USA) on the posterior side of the right arm midpoint (mm), three times, considering the average value between the three measures. The arm muscle circumference (AMC) was calculated according to the following equation [10]:

AMC(cm) = AC(cm) - [TS (mm) x 0,314]

The nutritional status was evaluated by three indicators: BMI/age percentile, height/age percentile and AMC value. The use of these three indicators provides more accurate information on nutritional status, as each reflects different body characteristics, such as longitudinal growth, adiposity and muscle mass. Malnutrition was defined according to 3 different criteria, according to Turcks at al (2016) [11]:

i. According to the percentile of the BMI/age: when p< 10.

ii. According to the percentile of height/age: when P < 10.

iii. By the AMC percentile value: when P ≤5.

Functional capacity was assessed through the 6-minute walk test (6MWT), following the the American Thoracic Society (ATS) standards, in a 30 meter corridor where participants were instructed to walk as fast as possible during the six minutes [12].

The HGS was obtained through the dynamometry, following the American Society of Hand Therapist (ASHT), using the Jamar ® dynamometer and the value used was the average of the 3 measurements [13]. Respiratory muscle strength was quantified through maximal inspiratory pressure (MIP) and maximal expiratory pressure (MEP), measurements performed by manovacuometry with a Wika ® analog manovacumeter in accordance with ATS standards [14].

Data were checked for normality by the Kolmogorov-Smirnov test. Parametric variables were presented by mean and standard deviation and non-parametric variables, such as median and minimum-maximum. Categorical variables were described using absolute and percentage frequencies. Student's T unpaired and Mann-Whitney tests were used for comparing two groups, according to the distribution of the data, parametric or not parametric, respectively. The comparison between 3 or more groups was performed by one-way analysis of variance (ANOVA) followed by Least Significant Difference test. The Chi-squared test was used to verify the existence of differences between proportions. Variables data of functional capacity and muscle strength were analyzed by the predicted value according to the formulas present in the literature [15-17].

Results

A total of 57 patients were evaluated with age of 13.26 ±3.1 Years, 42.1% of the males, 43.9% with mild obstructive disorder. Regarding the sum of the scores of Shwachman-Kulczycki clinical score, the patients reached a median of 70 (35-75) for the total of 75 points (Table 1).

Evaluating BMI/age, 59.6% of patients presented nutritional risk and 22.8% were considered malnourished. In relation to height/age, 15.8% presented malnutrition and according to AMC, 66.7% of the sample showed nourishment (Table 1).

Table 1: Demographic, clinic and nutritional description, from children and adolescents with cystic fibrosis, according to the genre.

	Total (n = 57)	Boys (n = 24)	Girls (n = 33)	p-Value
Age (years)	13,26 ± 3,1	13 ± 2,8	13,4 ± 3,4	0,631
Weight (kg)	41 ± 13,1	40,5 ± 11,2	41,3 ± 14,4	0,826
Height (cm)	149.3 ± 14,6	151,5 ± 15,3	147,8 ± 14,1	0,357
BMI(kg/m2)	17,9 ± 3,3	17,3 ± 2,2	18,3 ± 3,9	0,268
FEV1 (%)	83 (20 -109)	89 (20 -113)	79 (28-109)	0,077
FEV1 (Classes) (n/%)				
Grey 0	13 (22,8%)	9 (69,2%)	4 (30,8%)	0,267
Grey 1	25 (43,9%)	9 (36%)	16 (64%)	
Grey 2	4 (7%)	2 (50%)	2 (50%)	
Grey 3	8 (14%)	2 (25%0	6 (75%)	
Grey 4	5 (8,8%)	1 (20%)	4 (80%)	
Grey 5	2 (3,5%)	1 (50%)	1 (50%)	

Shwachman	70 (30 -75)	72 (30 -75)	70 (50 -75)	0,089
Genetic Mutation (N/%)				
No mapping	21 (36,8%)	11 (52,4%)	10 (47,6%)	0,519
F508del/F508del	8 (14%)	2 (25%)	6 (75%)	
F508del/Other	16 (28,1%)	7 (43,8%)	9 (56,2%)	
Other mutations	12 (21,1%)	4 (33,3%)	8 (66,7%)	
BMI/Age (Percentile)				
≥ 50 (n/%)	10 (17,5%)	5 (50%)	5 (50%)	0,844
≥ 10-< 50 (n/%)	34 (59,6%)	14 (41,2%)	20 (58,8%)	
< 10 (N/%)	13 (22.8%)	5 (38,5%)	8 (61,5%)	
Height/Age (Percentile)				
< 10 (n/%)	9 (15,8%)	2 (22,2%)	7 (77,8%)	0,188
≥ 10 (n/%)	48 (84,2%)	22 (45,8%)	26 (54,2%)	
AMC (Percentile)				
≤ 5 (n/%)	19 (33,3%)	11 (57,9%)	8 (42,1%)	0,088
> 5 (n/%)	38 (66,7%)	13 (34,2%)	25 (65,8%)	

FEV1: First Second Forced Exhalation Volume; PFE: Peak Exhalational Flow; BMI: Body Mass Index; AMC: Arm Muscle Circumference

Evaluating functional capacity, patients walked on average 634.7 ± 66,8m or 96.9% ± 9.5% of predicted value. Regarding the HGS, the patients reached 21.4 ± 8,9Kgf in dinamometry, which corresponds to 81.6 ± 17.8% of the predicted. The measurements of respiratory muscle strength demonstrated that patients had mean values of MIP and MEP of -82.3 ± 36,1cmH_2O and 71.5 ± 31,4cmH_2O, reaching 83.1 ± 35.3% and 61.1 ± 24.9% of the predicted, respectively. There was no statistically significant difference between boys and girls in the pneumofunctional evaluation, except for the absolute distance covered. However, after considering the influence of several factors using the prediction formula, it was noted that the genera obtained similar predicted values.

When comparing the means of distance covered in the 6MWT, HGS, MIP and MEP, according to the categories of BMI/age, there was a statistically significant difference between the distances traveled at the 6MWT by nourished (BMI/age ≥ 50) and malnourished (BMI/age < 10) and between the mean HGS values when compared to the malnourished with the nourished and the patients at risk nutritional risk with nourished (Table 2).

Table 2: Comparison of mean variables of interest, according to the categories of the percentiles of BMI/age.

	BMI/I (Percentile)			
	< 10	**≥10 - <50**	**≥ 50**	**p-Value**
6MWT (% predicted)	88 ± 19#	95,5 ± 8,3	100,9 ± 9,2#	0,027
Dynamometry (% predicted)	76,5 ± 12,8 #	78,9 ± 16,8 ##	92,8 ± 19,9 # ##	0,03
MIP (% predicted)	71,7 ± 45,7	84,7 ± 35,6	87,6 ± 24,9	0,522
MEP(% predicted)	58,9 ± 31,6	59,3 ± 24,7	67,4 ± 20,1	0,586

#p < 0.05 when compared to groups of BMI/age with percentiles ≤ 10 and ≥ 50; ## p < 0.05 when compared the groups of BMI/age between ≥ 10 and < 50 and ≥ p50.

BMI: Body Mass Index; 6MWT: 6-Minute Walk Test; MIP: Maximum Inspiratory Pressure; MEP: Maximum Expiratory Pressure

When comparing the means of the variables studied according to the height/age percentile, there was no statistically significant difference between the nourished and malnourished patients (Table 3). Still comparing the same variables, however, according to the classification of the percentile of AMC, only dynamometry presented a statistically significant difference between the nourished and malnourished (Table 4).

Table 3: Comparison of the means of the variables of interest, according to the categories of the percentiles of the height/age.

	Height/Age (Percentile)		
	≤ 10	> 10	p-Value
6MWT (% predicted)	98,8 ± 7,2	94,8 ± 12,2	0,342
Dynamometry (% predicted)	71,9 ± 12,6	83,5 ± 18,1	0,073
MIP (% predicted)	87,7 ± 43,6	82,2 ± 34	0,668
MEP (% predicted)	63,6 ± 25,5	60,6 ± 25	0,747

6MWT: 6-minute walk test; MIP: Maximum inspiratory pressure; MEP: Maximum expirat ory pressure

Table 4: Comparison of the means of the variables of interest, according to the categories of the percentiles of AMC/age.

	AMC (Percentile)		
	≤ 5	> 5	p-Value
6MWT (% predicted)	92,3 ± 14,8	96,9 ± 9,4	0,159
Dynamometry (% predicted)	74,8 ± 15,2	85,1 ± 18,1	0,038*
MIP (% predicted)	74 ± 37,2	87,6 ± 33,8	0,170
MEP (% predicted)	62,8 ± 23,1	60,2 ± 26	0,713

AMC: Arm Muscle Circumference; 6MWT: 6-Minute Walk Test; MIP: Maximum Inspiratory Pressure; MEP: Maximum Expiratory Pressure

Discussion

According to the results of this research, the impairment of nutritional status when assessed by the BMI/age index was associated with the decreased functional capacity and upper limb muscle strength in children and adolescents with CF.

In this study, it was demonstrated that most patients had good clinical conditions due to the high prevalence of mild obstructive ventilatory disorder and excellent Shwachman-Kulczycki score, which may explain the adequate functional capacity values, that is, distance travelled of 96.9 ± 9.5% of value predicted in 6MWT. A similar result was obtained by Gulmans et al. [18], who, when evaluating the reproducibility of the 6MWT in 23 children and adolescents with CF, found high values related to the distance covered in a sample that also had little impairment of pulmonary function [18].

Regarding the nutritional status, the prevalence of malnutrition was discrepant according to AMC and BMI/age evaluation. According to these variables, 33.3% and 22.8% of the patients were considered malnourished respectively. A similar result was found in the study conducted by Aquino and Cols (2014) that studied 46 Brazilian children and adolescents and observed that the prevalence of malnutrition was also different depending on the parameters used in nutritional status evaluation, with predominance of this when assessed by AMC [19].

Thus, the analysis of nutritional status by different indicators is Important for nutritional assessment and can detect malnutrition earlier because it evaluates growth, compartment composition and depletion of fat free mass (FFM) [20,21].

In this study, it was shown that patients with BMI/age lower than the 10th percentile presented a shorter distance traveled in the 6MWT, evidencing functional impairment in those considered malnourished. Pastré et al (2014), when evaluating 102 adults with CF in French reference centers, also concluded that functional capacity was correlated to BMI [22].

In the present study, patients who presented a compromised nutritional status by AMC (lower percentile equal to 5) and by BMI/age (percentile less than 10) have also presented a reduced HGS values. De Meer et al (1999) presented the same result in their study, in which lower values of AMC and FFM were associated with reducing peripheral muscle function, leading to an impact on functional capacity, even with adequate spirometry. However, functional capacity evaluation was performed by cycloergometry, which is a maximum test [23]. The difference in HGS between nourished and malnourished, according to AMC, reflects the decrease in arm muscle mass and overall muscle strength.

Meanwhile, Hallin et al (2011), when studying patients with COPD, found that the HGS and functional capacity values were associated with AMC and FFM, but not to BMI. This discrepancy may be associated with the age of the group with COPD that was composed of adults with a mean age of 64 years [24].

It is noteworthy that, with the improvement of therapies, the survival of these patients has increased in the decades. Thus, the improvement of functional capacity, muscle strength and nutritional status of children and adolescents will reflect in a better quality of life in adulthood.

According to the results of this study, a good nutritional status is important for the maintenance of both muscle strength and functional capacity. In this way, data suggests the importance of nutritional status in functional capacity and consequently in daily activities and quality of life of the CF patients.

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Conflict of Interest

We declare that financial interest and conflict of interest don´t exist.

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