Nutritional, clinical and socioeconomic profile of patients with cystic fibrosis treated at a referral center in northeastern Brazil*

Perfil nutricional, clínico e socioeconômico de pacientes com fibrose cística atendidos em um centro de referência no nordeste do Brasil

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Abstract

Objective: To describe the profile of patients with cystic fibrosis (CF). **Methods:** A prospective, cross-sectional study involving CF patients ≤ 18 years of age, evaluated between March and July of 2006 at a referral center in northeastern Brazil. Nutritional assessment was performed using Z scores for height/age (H/A), weight/age (W/A) and weight/height (W/H), as well as %weight/height (%W/H) and body composition measurements. Socioeconomic and clinical data were obtained. **Results:** Twenty-one patients were evaluated, 12 (57.1%) of whom were female. Mean age at diagnosis was 3.8 ± 3.9 years. The principal features at diagnosis were respiratory infection (85.7%), steatorrhea (66.7%) and nutritional deficit (47.6%). The mean Z scores for W/A, H/A and W/H were 0.73 ± 0.28, 0.34 ± 0.21 and 0.73 ± 0.35, respectively. Mean %W/H was 94.52 ± 1.58. The percentage of malnourished children assessed by Z score differed from that assessed by %W/H (nutritional deficit in 66.7% and 33.3%, respectively; $p > 0.05$). Socioeconomic status, clinical status and Shwachman score were better among well-nourished patients than among those classified as malnourished ($p < 0.05$ for Shwachman score). **Conclusions:** Normal nutritional status was identified based on nutritional indicators (W/A, H/A and W/H), whereas nutritional deficit was identified by assessing body composition. Socioeconomic factors proved favorable, especially maternal education and per capita income. Age at diagnosis was higher than that reported in the literature, although the Shwachman score and the incidence of respiratory infections demonstrated that the patients presented good clinical status.

Keywords: Cystic fibrosis; Nutritional assessment; Symptoms, clinical; Socioeconomic factors.

Resumo

Objetivo: Descrever o perfil de pacientes portadores de fibrose cística (FC). **Métodos:** Estudo transversal, prospectivo, avaliando fibrocísticos de ≤ 18 anos, durante o período de março a julho de 2006, em um centro de referência no nordeste do Brasil. A avaliação nutricional foi realizada pelo escore Z de altura/idade (A/I), peso/idade (P/I) e peso/altura (P/A) e %peso/altura (%P/A), além de medidas de composição corporal. Foram obtidos dados socioeconômicos e clínicos. **Resultados:** Foram avaliados 21 pacientes, sendo 12 (57,1%) do sexo feminino. A média de idade de diagnóstico foi de 3,8 ± 3,9 anos, e as principais características ao diagnóstico foram infecção respiratória (85,7%), esteatorreia (66,7%) e déficit nutricional (47,6%). A média de escore Z para P/I, A/I e P/A, respectivamente, foi de −0,73 ± 0,28, −0,34 ± 0,21 e −0,73 ± 0,35. A média de %P/A foi de 94,52 ± 1,58. O percentual de desnutridos divergiu quando avaliado pelo escore Z e %P/A (déficit nutricional em 66,7% e 33,3%, respectivamente; $p > 0,05$). Os pacientes eutróficos apresentaram melhores condições socioeconômicas ($p > 0,05$) e clínicas, com melhor escore de Shwachman ($p < 0,05$) quando comparados aos distróficos. **Conclusões:** Eutrofia foi encontrada através dos indicadores nutricionais (P/I, A/I e P/A), ao passo que déficit nutricional foi encontrado quando avaliada a composição corporal. As condições socioeconômicas apresentaram-se favoráveis, principalmente em relação ao grau de instrução materna e renda per capita. A idade de diagnóstico foi maior do que o relatado na literatura, embora o escore de Shwachman e o número de infecções respiratórias tenham demonstrado que os pacientes tinham boas condições clínicas.

Descritores: Fibrose cística; Avaliação nutricional; Sintomas clínicos; Condições socioeconômicas.

Introduction

Cystic fibrosis (CF) is an autosomal recessive genetic disease that affects various systems of the human body, being characterized by a generalized dysfunction of the exocrine cells and resulting in COPD, with an accumulation of thick and purulent secretion, recurrent respiratory infections, progressive loss of pulmonary function, decreased mucociliary clearance, pancreatic insufficiency and increased concentrations of sweat electrolytes, as well as impairing, albeit less often, the intestine, liver, biliary tract and genital tract.[1,2]

It is estimated that the prevalence of CF in Brazil is 1/10,000 live births, although there are variations between geographic regions in terms of the frequency of mutations. In the southern region of Brazil, this prevalence is similar to that reported for the Caucasian population of central Europe (from 1/2,000 to 1/5,000 live births).[1,3,4]

In recent decades, knowledge regarding genetic mutation and the development of diagnostic techniques that have allowed progressively earlier diagnosis have greatly improved the prognosis and the quality of life of CF patients. However, the prevalence of malnourishment and growth retardation in these patients continues to be significant. In the USA, approximately 20% of children and teenagers with CF present weight and height below the 5th percentile.[5]

The intimate relationship that changes in body composition and malnutrition have with pulmonary function is well established in the literature.[1] Malnutrition negatively affects the function of respiratory muscles, lung elasticity and immune response, since it increases the risk of pulmonary colonizations, especially when accompanied by a decrease in lean body mass.[6]

The problems of malnutrition presented by CF patients are multifactorial and can be attributed to the energy imbalance caused by chronic respiratory disease, poor nutrient absorption caused by pancreatic insufficiency, changes in enterohepatic circulation of biliary salts and antibiotic therapy, as well as to the possibility of being aggravated by anorexia caused by gastroesophageal reflux or cough, increased metabolism, especially in the presence of infections, and psychosocial stress.[7,8]

The objective of this study was to describe the nutritional, clinical and socioeconomic profile of CF patients seen at a referral center in northeastern Brazil, given the scarcity of studies evaluating such conditions in patients living in this particular region.

Methods

We conducted a prospective, cross-sectional study developed in the pediatric department of the *Instituto Materno Infantil Professor Fernando Figueira* (IMIP, Professor Fernando Figueira Mother and Child Institute), between March and July of 2006. Participants were recruited from among children and adolescents who sought treatment in the department during the study period. Those ≤ 18 of years of age with a confirmed diagnosis of CF were included. None were excluded.

The diagnosis of CF was made based on a finding of at least one of the clinical manifestations characteristic of the disease, as well as on at least two determinations of chlorite counts in sweat, by iontophoresis with pilocarpine, presenting values above 60 mEq/L, or on the presence of two known CF mutations identified through genetic studies.[9]

For the socioeconomic evaluation, we used a structured questionnaire designed to collect data related to maternal identity (age, level of education and parity), number of residents in the household and monthly per capita income.
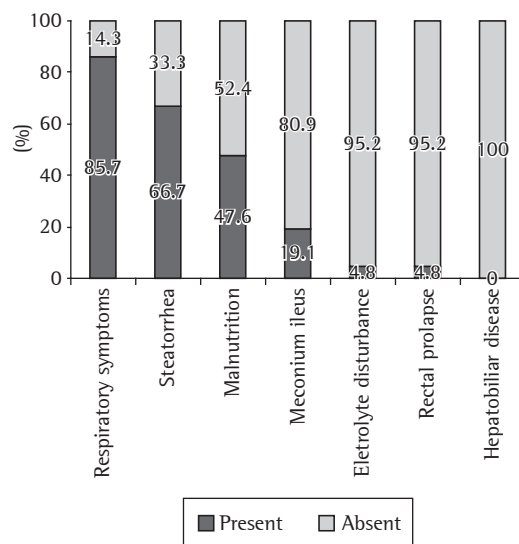
Figure 1 – Clinical manifestations present at diagnosis in patients with cystic fibrosis treated at a referral center in Recife, Brazil.

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Based on the city of residence, patients were divided into three groups by region: the greater metropolitan region of Recife; the countryside of the state of Pernambuco; and other states.

Data were collected regarding the following: gestational age of the patients at birth, defined as pre-term (< 37 weeks) or term (≥ 37 weeks); birth weight, defined as underweight (< 2500 g) or appropriate (≥ 2500 g); and the duration of exclusive breastfeeding.

In the clinical evaluation, the age at diagnosis and clinical manifestations present at diagnosis were noted. The evaluation of clinical severity of the disease was based on the Shwachman score, applied by a pediatric pulmonologist, who evaluated, in points of equal weights, the following items: general activity, pulmonary profile, radiological findings and nutritional state. Each criterion is scored from 5 to 25 points, lower scores indicating greater severity.[10] Pulmonary colonization was defined as presenting a positive culture for *Pseudomonas aeruginosa*, *Staphylococcus aureus* (oxacillin-sensitive or -resistant) or *Burkholderia cepacia*.

For the assessment of the nutritional status, the individuals were divided into three groups according to age: under 5 years of age; between 5 and 10 years of age; and above 10 years of age. Measurements of weight, height, arm circumference (AC) and triceps skinfold thickness (TST) were used. For children under 2 years of age, we used a digital scale (Filizola, Indústria Filizola S/A, São Paulo, Brazil) with a 15-kg capacity and precision to within 100 g, as well as an infantometer for measuring length. For children

over 2 years, we used a digital scale (Filizola®, Indústria Filizola S/A, São Paulo, Brazil) with a 150-kg capacity and precision to within 50 g, as well as a stadiometer for height measurement.

These measurements were expressed in Z scores for weight/age (W/A), height/age (H/A) and weight/height (W/H) based on age and gender, comparing them with the standards of the World Health Organization (WHO),[11] for individuals under 5 years of age, and of the National Center for Health Statistics,[12] for those above 5 years of age. Those presenting a Z score < –1 for any evaluated indicator were considered to have nutritional deficit, whereas those presenting a Z score ≥ –1 SD were considered to present normal nutritional status. The percentage of ideal weight in relation to height (%W/H) was also calculated according to the recommendations of the US Cystic Fibrosis Foundation Consensus Report, which classify < 90% as being at nutritional risk and ≥ 90% as presenting normal nutrition.[13,14]

For AC measurement, a non-elastic metric tape was used. For TST measurement, we used a Lange Skinfold Caliper (Cambridge Scientific Industries, Cambridge, MD), designed for use with children over 6 years of age. Measurements were conducted at the midpoint between the olecranon and the acromion of the non-dominant arm of the patient.

Muscle and fat mass was evaluated by calculating the arm muscle area (AMA) and arm fat area (AFA) based on the AC and TST, respectively, using the parameters established by Frisancho.[15] The analysis was conducted using

Table 1 – Anthropometric data and Shwachman score by age bracket, expressed in mean and SD, in patients with cystic fibrosis treated at a referral center in Recife, Brazil.

Parameter	Age (years)			p*
	0 ⊢ 5	5 ⊢ 10	10 ⊢ 18	
	(n = 7)	(n = 6)	(n = 8)	
Indicator				
W/A	–0.73 ± 1.26	–0.75 ± 1.00	–0.89 ± 1.13	NS
H/A	–0.18 ± 1.71	–0.39 ± 0.93	–0.52 ± 1.56	NS
W/H[a]	–0.75 ± 0.96	–0.74 ± 0.90	–	NS
%W/H	95.14 ± 5.83	95.97 ± 8.37	92.68 ± 21.41	NS
AMA%[b]	–	35.41 ± 12.33	64.30 ± 16.34	0.013
AFA%[b]	–	49.66 ± 17.76	59.97 ± 36.48	NS
Shwachman score	81.67 ± 87.51	82.50 ± 16.05	81.43 ± 11.44	NS

W/A: weight/age; H/A: height/age; W/H: weight/height; %W/H: %ideal weight/height; AMA: arm muscle area; AFA: arm fat area; NS: not significant. [a]Measurements taken in children under 10 years of age; [b]Measurements taken in children under 5 years of age. *One-way ANOVA.

percentage adjustment in relation to the 50th percentile, defining patients with adjustment < 90% as having nutritional deficit, and those with adjustment ≥ 90% as presenting normal nutritional status.

For the assessment of nutritional indicators, the WHO Anthro 2005 program (WHO, Geneva, Switzerland) was used.[16] We used the Kolmogorov-Smirnov normality test, and the distribution was found to be symmetric, the data being presented as mean and standard deviation. The Student's t-test was used for comparing two independent groups and one-way ANOVA was applied for more than two groups. Fisher's exact test was used for comparing proportions. A significance level of 0.05 was adopted.

The study was approved by the Ethics in Research Committee of the IMIP and was later clarified in detail to the parents or legal guardians, who gave written informed consent authorizing the participation of each patient in the study.

Results

Data regarding 21 individuals were collected. Of those, 12 (57.1%) were female and 9 (42.9%) were male, with a mean age of 8.4 ± 4.6 years. Of the 21 participants, 7 (33.3%) were under 5 years of age, 6 (28.6%) were 5-10 years of age, and 8 (38.1%) were over 10 years of age.

As for the place of origin, 11 (52.3%) of the participants were residents of the metropolitan region of Recife, 9 (42.9%) were residents of Pernambuco, and 1 (4.8%) was a resident of another state (Paraíba).

Age at diagnosis ranged from the neonatal period to 14 years (mean, 3.8 ± 3.9 years). In 2 cases (9.5%), the diagnosis was made after 10 years of age. The principal clinical manifestations present at diagnosis were: acute or persistent respiratory symptoms (85.7%), steatorrhea (66.7%) and nutritional deficit (47.6%; Figure 1). Four patients (19.1%) had a family history of CF.

The mean scores for the nutritional indicators (W/A, H/A and W/H) in each age bracket were found to be within the limit of normality, which was defined as a Z > −1 for each of the parameters evaluated. In general, the indicators of nutritional status were better among those below 5 years of age than among those in the other age brackets. In terms of %W/H, the impairment of nutritional status was greater in individuals older than 10 years of age. However, there was no statistically significant difference among the age brackets for either indicator (p > 0.5; Table 1).

Our results regarding body composition differ from those presented by the anthropometric indicators. There was a marked deficit regarding lean body mass, represented by AMA, as well as regarding the fat compartment, represented by AFA, with mean scores below 60% of adjustment for normal nutritional status, showing severe depletion of body reserves and being more pronounced in the 5-10 year age bracket, with a statistically significant difference for AMA (Table 1).

The percentage of well-nourished and malnourished individuals varied according to the parameter used for the classification of the nutritional status. Using the parameters of the WHO (Z score),[11,12] 66.7% of the individuals were classified as malnourished, whereas when the same were evaluated by the recommendation of nutrition consensuses in CF (%W/H),[13,14] only 33.3% were classified as malnourished. In the interest of identifying significant differences between the classification of malnutrition (nutritional deficit) and normal nutritional status, Fisher's exact test was applied (p > 0.05; Table 2).

When nutritional state was evaluated according to age at diagnosis, the nutritional indicators were found to be better among those diagnosed before the age of 1 year than among those diagnosed at a later age, although there were no statistically significant differences.

According to the Shwachman score, 52.6% and 47.4% of the patients presented the mild and moderate forms of the disease, respectively. Based on this score, none of the participants presented the severe form of the disease. Three patients (14.3%), all of whom were younger than 10 years of age, presented excellent clas-

Table 2 – Classification of nutritional status, according to the World Health Organization and nutritional consensuses on CF, in patients with cystic fibrosis treated at a referral center in Recife, Brazil.

Nutritional status	WHO, n (%)	Consensuses, n (%)
Normal nutrition	7 (33.3)	14 (66.7)
Nutritional deficit	14 (66.7)	7 (33.3)
Total	21 (100)	21 (100)

WHO: World Health Organization. Fisher's exact test (p > 0.05).

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Table 3 – Neonatal, socioeconomic and clinical characteristics, according to the nutritional status of patients with cystic fibrosis treated at a referral center in Recife, Brazil.

Characteristic	Normal nutrition[a] (n = 7)	Nutritional deficit[b] (n = 14)	p
Age (years)	6.94 ± 5.11	8.64 ± 4.45	NS*
Neonatal characteristics			
Birth weight (g)	3.139 ± 512	3.351 ± 903	NS*
Exclusive breastfeeding period (months)	3.33 ± 1.16	2.62 ± 2.47	NS*
Socioeconomic characteristics			
Mother's age (years)	36.50 ± 7.94	31.92 ± 7.35	NS*
Maternal education (years)	11.33 ± 2.42	9.00 ± 4.73	NS*
Number of children	2.86 ± 1.22	2.31 ± 0.86	NS*
Number of residents in the household	5.00 ± 2.31	4.31 ± 1.11	NS*
Per capita income (R$)	407.88 ± 395.15	226.46 ± 199.48	NS*
Clinical characteristics			
Age at diagnosis (years)	2.44 ± 3.48	4.52 ± 3.98	NS*
Shwachman score	94.29 ± 5.35	74.58 ± 12.52	0.001*
Colonization with *aeruginosa*	28.6%	71.4%	NS**
Colonization with oxacillin-sensitive *aureus*	33.3%	66.7%	NS**
Colonization with oxacillin-resistant *S. aureus*	33.3%	66.7%	NS**
Colonization with *cepacia*	25.0%	75.0%	NS**

NS: not significant. [a]Weight/Age, Height/Age, Weight/Height ≥ −1 SD; [b]Weight/Age, Height/Age, Weight/Height < −1 SD. *Student's t-test; **Fisher's exact test.

sification (100 points), indicating that clinical status is better in younger children, although there were no statistically significant differences among the age brackets (Table 1).

Neonatal, socioeconomic and clinical characteristics were evaluated according to nutritional status (well-nourished *vs.* malnourished), based on the Z score. These characteristics can be seen in Table 3. Although there were no statistically significant differences among the groups, longer duration of exclusive breastfeeding period, older maternal age, higher maternal level of education and higher per capita income were all found to improve nutritional status. As for the clinical characteristics, individuals diagnosed at an earlier age more often presented normal nutritional status and less often presented pulmonary colonization with any of the microorganisms evaluated. The mean Shwachman score was significantly higher in patients with normal nutritional status, showing the influence that nutritional status has on this index.

Discussion

There are few data in the literature regarding socioeconomic data of individuals with CF, which makes it impossible to compare our results with the social situations in other Brazilian localities. Despite this fact, we emphasize the importance of the study of these variables as a means of expanding knowledge regarding the specific social aspects of this population.

The better nutritional status found in our sample can be explained by the favorable neonatal and socioeconomic conditions.

Birth weight and breastfeeding are important predictors of nutritional status. Gestational age and appropriate birth weight, together with longer duration of breastfeeding, act as protective factors against the occurrence of frequent illnesses, especially those of infectious origin, and malnutrition,[17] which might have contributed to the fact that the nutritional status presented by the individuals studied was better than that reported in other studies.[18-20]

In relation to maternal education, more years of schooling favors better nutritional status, as well as improving patient adherence to treatment. Family income can be considered an important determinant of health conditions by the influence it exerts on the possibility of acquiring and using goods and services essential to the maintenance of the health status, such as food, housing, clothing and sanitation.[21]

The mean age at diagnosis in our sample was similar to that found in a study conducted in southeastern Brazil.[22] However, it has been reported that the median age at diagnosis in the USA is 6 months.[23] In our study, patients with normal nutritional status presented a lower mean age at diagnosis than did those considered malnourished, showing the need for an early diagnosis, especially with the implementation of neonatal testing, which is still rarely conducted in our region. Early diagnosis is important for prophylaxis of pulmonary infections and replacement of pancreatic enzymes in cases of pancreatic insufficiency, improving the absorption of nutrients with the objective of maintaining good nutritional status, decreasing complications resulting from the disease and increasing survival.[24]

Clinical manifestations can vary greatly and can appear early or late. The clinical manifestations reported to be most common to a diagnosis of CF are persistent cough, chronic diarrhea and malnourishment,[22,25,26] as was observed in our study. The Shwachman score showed favorable clinical conditions in our sample, in which the mean score was 81.8 ± 14.2, compared with presented means of 74 ± 17[26] and 56 ± 15 in two other studies,[26,27] the latter of which involved hospitalized patients presenting greater clinical impairment.

In national[19,20] and international[5,18] studies of CF patients, some degree of malnutrition, as well as greater impairment of nutritional status in those over 10 years of age, has been described. Those findings were attributed to greater nutritional demand, due to the rapid growth or increase in energy expenditure related to pulmonary infections, which are more common in CF patients, as well as to changes in endocrine function, due to a higher probability of developing diabetes in this age bracket.[28] There can also be lower adherence to enzyme reposition therapy, changes in diet and increased physical activity in this age bracket.[20] It is of note that the individuals studied presented pancreatic insufficiency and made appropriate and regular use of pancreatic enzymes. None of them had been diagnosed with diabetes mellitus.

In CF patients, the loss of body mass normally occurs in the body fat and lean body mass compartments, which might be associated with a significant increase in the catabolism of proteins and fat reserve. Energy balance and protein metabolism are adversely affected by episodes of acute exacerbation of pulmonary infections, which impairs the function of respiratory muscles, exercise tolerance and immune function, thereby worsening the prognosis of these patients.[20,29]

When we evaluated nutritional status, our findings varied depending on whether we applied the standards proposed by the WHO or those recommended by the international consensuses on CF. In addition, the evaluation of body composition revealed that few of the individuals studied were considered well-nourished, and that there were many cases of malnutrition. This was also reported in another study.[30]

We can conclude that the nutritional status of the patients treated at the center in question varied widely according to the parameters used for classification, with a tendency toward normal nutritional status regarding anthropometric indicators (weight and height) and toward nutritional deficit regarding the muscle and fat compartments, showing the need for a specific and more detailed nutritional assessment which would include all of these indicators. Neonatal and socioeconomic conditions were favorable, principally in relation to maternal level of education and per capita income, which directly contribute to a higher adherence to treatment, as well as to better nutritional status. In relation to clinical conditions, individuals with better nutritional status presented younger age at diagnosis, fewer pulmonary infections and better Shwachman scores.

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