Results of nutritional intervention in children and adolescents with cystic fibrosis

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Abstract

Objective: Few studies have verified longitudinally the evolution of the nutritional status of patients with cystic fibrosis. The objective of this study is to follow the evolution of the nutritional status, body composition and energy consumption, macronutrients and micronutrients ingested by children and adolescents by means of nutritional interventions at the Clinic of Cystic Fibrosis/Pediatric Pneumology of the Department of Pediatrics of Universidade Federal de São Paulo.

Methods: 18 patients were involved in this study, thirteen males and five females with ages ranging from 0.3 to 18.4 years. We performed three evaluations: evaluation 1 (M1- prenutritional intervention), M2 after 6 months, and M3 after 12 months. In these three instances we verified: the z score for weight/age, weight/height and height/age and the calculation of a 3-day diet record. We verified the body composition (anthropometry) in M1 and M3. The nutritional interventions were hypercaloric, hyperproteic, with adequate amount of ingested macronutrients and micronutrients.

Results: We observed an increase in the z score for height/age (M1=-1.07; M2=-0.69; M3=-0.50) and fat-free mass after the nutritional interventions, without improvement in the z score for weight/height and fat mass. We verified an increase in the energy intake during M2 (139%) and M3 (132%) compared to M1 (106%). Remarkable increase in the intake of protein, calcium, iron and vitamin C by the patients was found. The occurrence of anemia was found in 44% (8/18) of the patients.

Conclusion: The improvement of the z score in height/age and fat-free mass was probably due to the increase in energy consumption after the nutritional intervention. A significant improvement in the z score for weight/height and fat mass was not found, probably due to a gain in height and fat-free mass.


Introduction

Cystic fibrosis is characterized by a dysfunction of general exocrine glands, with production of thick and abnormal mucus, which may obstruct the ducts of several affected organs, causing chronic lung disease, pancreatic insufficiency, male infertility and high concentration of sodium and chloride in the sweat.1
In spite of recently accomplished advances regarding clinical and nutritional treatment, the incidence of malnutrition and growth deficit among patients with cystic fibrosis is still significant. The Cystic Fibrosis Foundation from the United States pointed out that, in 1997, 24% of the patients were below the 5th percentile for weight and 19% were below the 5th percentile for height. Pulmonary function and nutritional status are closely related, since significant weight loss may lead to the reduction of lean body mass, with consequences to respiratory muscles and pulmonary elasticity. Therefore, protein-energy malnutrition is considered the worst prognostic factor of this disease.

Patients are affected by malnutrition and growth deficit because they have difficulty obtaining their energy supply and due to energy loss caused by the disease, which is a consequence of chronic pulmonary inflammations and infections, malabsorption caused by pancreatic insufficiency, alteration in the enterohepatic circulation of bile salts and antibiotic therapy. The presence of anorexia, which is a result of gastroesophageal reflux and/or cough, chronic respiratory infections and psychosocial stress, may worsen malnutrition. The diet therapy is responsible for improving the patient’s nutritional status or keeping it appropriate, and breaking the malnutrition-infection vicious circle.

The main purpose of this study was to assess the impact of nutritional intervention in patients with cystic fibrosis, both pancreatic-insufficient and pancreatic-sufficient patients, by means of an anthropometric examination and body composition assessment in a longitudinal study.

Methods

The subjects of this study were recruited at the Cystic Fibrosis Clinic of the Center of Pediatric Pneumology, Escola Paulista de Medicina/Universidade Federal de São Paulo. The study involved 18 patients who attended at least four visits in 12 months. The initial age of follow-up ranged between 0.3 and 18.4 years; 13 of these patients were males and five were females. Patients were assessed by a multiprofessional team, which included a nutritionist, pediatricians (specialized in pulmonary and gastric diseases) and a physical therapist.

The diagnosis of cystic fibrosis was accomplished based on the presence of at least one of the clinical symptoms that characterize the disease. It was confirmed through the ionic analysis of the sweat, with two positive samples of sodium and chloride at least, and through the detection of the disease gene by means of a genetic test.

The study included data collected from January 1998 to November 2000. The evaluations were performed at three different moments. The initial moment (M1) was before the nutritional intervention, the second moment (M2) was after a period of 6 months, and moment 3 (M3) occurred after a period of 12 months (final evaluation). The research protocol was approved by the Medical Ethics Committee of Universidade Federal de São Paulo. Before the study began, the guardians signed a term of agreement, allowing patients’ participation in the research.

Data collected retrospectively through medical records

Shwachman score: performed by pediatricians specialized in pulmonary diseases at the initial moment (M1) and at the final moment (M3) of nutritional follow-up. It is considered a trustful method for the evaluation of cystic fibrosis severity. It evaluates the following items, according to points with equivalent value: general activity, data from the physical examination related to the pulmonary condition, radiological findings and nutritional status. Even though there are other tests designed for the clinical evaluation of these patients, Shwachman score is the method chosen by the service.

Lung colonization: the presence of colonization with Pseudomonas aeruginosa and Staphylococcus aureus was detected. The persistence of the bacterium presence for a period longer than six months was considered chronic colonization. Patients that presented chronic colonization with P. aeruginosa were treated with antibiotics every three months, for at least 14 days, alternating the treatment with hospital or clinic admission.

Admissions: during the follow-up period, data regarding the causes of admissions were collected.

Hemoglobin and hematocrit: were collected at the beginning of the follow-up period, based on the recommendations of World Health Organization for cutoff points.

Data collected prospectively

Anthropometry

Weight and height were measured at three moments according to the techniques mentioned by Frisano.

– Weight: children up to two years old were weighed using pediatric digital scales, and children older than 2 years, were weighed by means of adult scales (of the brand Filizola). Children were barefoot and wore as few clothes as possible.

– Length/height: children up to two years old were measured with a wooden horizontal anthropometer, and children older than two, were measured with a wooden stadiometer, 200 cm long.

– Arm perimeter (AP) and tricipital skinfold thickness (TST): patients’ right arms were measured at M1 and M3, according to the techniques mentioned by Deurenberg et al. and by WHO. TST was measured with Lange skinfold caliper (Cambridge Scientific Industries, Cambridge, Md) and AP was obtained using a non-extensible tape measure.
Body composition

Based on AP and TST measures, we were able to assess fat body mass and muscle mass by calculating the arm muscle area (AMA), the arm fat area (AFA) and the percentage of body fat. Since there were no data related to patients younger than one year old in the table used as a reference, we measured the values for those patients, but we did not classify them.

Classification of the nutritional status

The data collected were assessed through the z score for weight/age, weight/height and height/age. The weight/height indicator was also expressed as an adequacy percentage, according to the median of the reference population, as recommended by the Cystic Fibrosis Consensus (ANTHRO, Software for Calculating Pediatric Anthropometry, v.1, 02, 1999). We adopted the anthropometric standard of reference of the National Center for Health Statistics.

Nutritional Intervention

The nutritional education sessions occurred once a month, on an individual basis, and for patients with stable clinical and nutritional aspects, it was performed once every two months.

Nutritional control

We recommended ingestion of energetic value from 120 to 150% of the RDAs (Recommended Dietary Allowances, 1989) for age and gender, and if necessary, 45% of the total value should consist of lipids, according to the regulations of the Cystic Fibrosis Consensus. We suggested the minimal adjustment for age of 80% of the RDAs for iron, calcium, zinc, vitamin *C* and 100% for protein. At the three different follow-up moments, we calculated a three-day record or, in case there were no records, we calculated the record of the usual food intake using the CIS/EPM software, which analyzed the proportion of macronutrients, micronutrients and food energy values.

Energetic oral supplements: recommended for patients with weight/height <85% or when there is a decrease in weight gain.

   a) Lipid supplement: with raw vegetal oil or medium-chain triglycerides, ingested during the main meals, with an initial mean dosage of 16g/day, which might be increased to 24g/day, if necessary.

   b) Dietetic supplement: ingested orally by means of manufactured supplement, mixed with water or milk.

Compliance with oral nutritional supplement: the daily intake of the recommended quantity during a minimal period of six months was considered to be compliant with the treatment.

Compliance with the pancreatic enzymes: in the nutritional follow-up visits, we reinforced the correct use of pancreatic enzymes and its importance to nutritional improvement. Those patients that did not correctly and sufficiently follow the intake of enzymes at any time during the follow-up were considered as noncompliants.

Statistical study

With the purpose of comparing the results of the three different moments for each patient according to the variables analyzed, we used: Friedman’s two-way analysis of variance, and when there was a significant difference, this analysis was complemented by the multiple comparison test. The Wilcoxon test was employed in order to compare the results observed at the initial moment (M1) and final moment (M3) with regard to the body composition of each patient. Cochran’s g-test assessed the adequacy (+) in the three periods of the study. In all tests the level for rejection of the null hypothesis was established at 0.05 or 5%, using an asterisk to indicate the significant values.

Results

During the follow-up period, 11 patients (61.1%) presented episodes of colonization with *S. aureus*, five (27.8%) with *P. aeruginosa* and another five (27.8%) with *P. aeruginosa crônica* (Figure 1).

Between periods M1 and M2, four patients were hospitalized, three of them were breastfed infants. Three patients were submitted to a decontamination of *P. aeruginosa*, and one breastfed infant had to be hospitalized twice because of bowel subobstruction and a surgery due to...
bowel bridle. Between periods M2 and M3, seven patients were hospitalized, six of them underwent decontamination of *P. aeruginosa* and, among them, two patients were hospitalized one more time during that period. The admissions were caused by bronchopneumonia and decontamination of *S. aureus*. One patient was hospitalized because of worsened pulmonary condition (Figure 1).

There was a significant increase in the values of Schwachman score at the end of the follow-up compared to the period prior to the intervention, and the score classification was not altered (Table 1).

### Table 1 - Patients with cystic fibrosis, according to the mean and median of Shwachman score at follow-up periods M1 (initial) and M3 (final). Result of Wilcoxon test

<table>
<thead>
<tr>
<th>Period</th>
<th>M1</th>
<th>M3</th>
<th>M1 x M3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean</td>
<td>6.8</td>
<td>2.8</td>
<td>T calc:13.5</td>
</tr>
<tr>
<td>Median</td>
<td>7.5</td>
<td>5.0</td>
<td>P&lt;0.02</td>
</tr>
</tbody>
</table>

We observed improvement in the height/age z score after the nutritional intervention during periods M2 and M3, compared to M1. However, there was almost no alteration in the weight/height z score and we did not detect significant improvement of the weight/age z score after the intervention (Table 2).

There was a significant increase in patients’ lean body mass, represented by AMA at M3 compared to M1, although there was no improvement of AFA and the percentage of fat body mass (Table 3).

There was a significant improvement of energy intake at M2, compared to M1. After analyzing micronutrients, we observed a significant improvement of the calcium intake at M2, compared to M1. We also observed a significant improvement of iron intake, but that improvement was not detected when we analyzed the number of patients who presented appropriate values of iron. We did not verify a significant improvement of the zinc and vitamin C intake, although there was a remarkable increase in the mean amount of vitamin C intake during M2 and M3, compared to M1 (Table 4 and 5).

### Discussion

Thousands of mutations related to the cystic fibrosis have already been described and the clinical manifestations of this disease present great variability. DF508 Mutation is the most frequent among Caucasian people, and when it occurs in homozygotes, it is strongly associated with pancreatic insufficiency. In this study, nine out of 11 patients with pancreatic insufficiency presented the DF508 mutation and, among the patients that did not present pancreatic insufficiency, only two patients had this mutation in one of the chromosomes. Regarding the manifestation of the lung disease, studies have not confirmed the association with the genotype.

### Table 2 - Patients with cystic fibrosis, according to the nutritional indicators during follow-up (M1, M2 and M3). Result of Friedman’s two-way analysis of variance

<table>
<thead>
<tr>
<th></th>
<th>M1</th>
<th>M2</th>
<th>M3</th>
<th>$\chi^2$ calculated</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>% W/H</td>
<td>94.35</td>
<td>96.65</td>
<td>93.90</td>
<td>3.44</td>
<td>SL</td>
</tr>
<tr>
<td>W/H z score</td>
<td>-0.83</td>
<td>-0.58</td>
<td>-0.80</td>
<td>1.36</td>
<td>SL</td>
</tr>
<tr>
<td>W/A z score</td>
<td>-1.36</td>
<td>-0.93</td>
<td>-0.90</td>
<td>4.00</td>
<td>SL</td>
</tr>
<tr>
<td>H/A z score</td>
<td>-1.07</td>
<td>-0.69</td>
<td>-0.50</td>
<td>11.37*</td>
<td>M1 &lt; M2 and M3</td>
</tr>
</tbody>
</table>

* P<0.01

### Table 3 - Mean and median of body composition of patients with cystic fibrosis analyzed through arm muscle area (AMA), arm fat area (AFA) and percentage of fat body mass at moments M1 (initial) and M3 (final). Result of Wilcoxon test analysis

<table>
<thead>
<tr>
<th></th>
<th>M1</th>
<th>M3</th>
<th>$z$ calculated</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>AMA</td>
<td>16.58</td>
<td>20.83</td>
<td>3.55*</td>
<td>M1 &lt; M3</td>
</tr>
<tr>
<td>AFA</td>
<td>6.48</td>
<td>7.47</td>
<td>1.81</td>
<td>SL</td>
</tr>
<tr>
<td>% Fat</td>
<td>27.57</td>
<td>26.07</td>
<td>1.54</td>
<td>SL</td>
</tr>
</tbody>
</table>

* P<0.001

* P<0.01
The data collected demonstrate the presence of homozygous DF508 genetic mutation in four (22.2%) patients, and seven (38.9%) presented heterozygous DF508 genetic mutation. Other studies performed in Brazil suggest the presence of this mutation among 27% and 53% of the patients. The occurrence of DF508 mutation varies according to the incidence of the Caucasian population, leading Raskin et al. to declare that the frequency of this mutation in Brazil is similar to the frequency in Southern Europe, due to the European immigration to Brazil during the colonization period.

P. aeruginosa is the main bacterium responsible for progressive pulmonary damage. The incidence of P. aeruginosa increases as patients grow older and it affects from 70 to 90% of patients. Some studies have detected weight loss when there is lung colonization with P. aeruginosa, however the mechanism that causes this loss still needs to be explained. In a study performed with rats, weight loss was also detected when there is acute lung colonization with P. aeruginosa, and this loss is correlated to the production of cytokines of the inflammatory state, and it is not associated with the alteration of the pulmonary response. In this study, we detected the presence of lung colonization with P. aeruginosa and S. aureus at all ages during the follow-up period, and in patients older than nine years old we verified greater occurrence of chronic colonization with P. aeruginosa (4/5) (Figure 1).

Table 4 - Patients with cystic fibrosis, according to the intake of energy and nutrients during follow-up (M1, M2 and M3). Result of Friedman’s two-way analysis of variance

<table>
<thead>
<tr>
<th></th>
<th>M1</th>
<th>M2</th>
<th>M3</th>
<th>$\chi^2$ calculated</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>% Energy/RDAs</td>
<td>106</td>
<td>139</td>
<td>132</td>
<td>9.00*</td>
<td>M1 &lt; M2</td>
</tr>
<tr>
<td>% Protein/RDAs</td>
<td>224</td>
<td>290</td>
<td>303</td>
<td>5.77</td>
<td>SL</td>
</tr>
<tr>
<td>% Lipids/TEV†</td>
<td>32.8</td>
<td>36.5</td>
<td>36.4</td>
<td>3.44</td>
<td>SL</td>
</tr>
<tr>
<td>% Carbohydrate/TEV†</td>
<td>55.3</td>
<td>51.0</td>
<td>50.5</td>
<td>2.11</td>
<td>SL</td>
</tr>
<tr>
<td>% Protein/TEV†</td>
<td>11.9</td>
<td>12.5</td>
<td>13.1</td>
<td>2.11</td>
<td>SL</td>
</tr>
<tr>
<td>% Calcium/RDAs</td>
<td>75.2</td>
<td>108.3</td>
<td>92.7</td>
<td>8.77*</td>
<td>M1 &lt; M2</td>
</tr>
<tr>
<td>% Zync/RDAs</td>
<td>89.8</td>
<td>90.8</td>
<td>99.4</td>
<td>0.25</td>
<td>SL</td>
</tr>
<tr>
<td>% Iron/RDAs</td>
<td>102.5</td>
<td>133.7</td>
<td>141.2</td>
<td>8.44*</td>
<td>M1 &lt; M2 and M3</td>
</tr>
<tr>
<td>% Vitamin C/RDAs</td>
<td>140.4</td>
<td>285.3</td>
<td>490.8</td>
<td>5.44</td>
<td>SL</td>
</tr>
</tbody>
</table>

* P<0.02 † TEV - total energy value

Table 5 - Patients with cystic fibrosis, according to the mean of adequacy percentage (+) of energy, calcium, zync, iron and vitamin C intake regarding the RDAs for age during the follow-up periods. Result of the analysis of Cochran’s g-test

<table>
<thead>
<tr>
<th></th>
<th>M1</th>
<th>M2</th>
<th>M3</th>
<th>g calculated</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adequacy/Energy n(+)</td>
<td>7</td>
<td>13</td>
<td>9</td>
<td>6.22*</td>
<td>M1 &lt; M2</td>
</tr>
<tr>
<td>%(+ )</td>
<td>38.9</td>
<td>72.2</td>
<td>50.0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Adequacy/Calcium n(+)</td>
<td>6</td>
<td>11</td>
<td>10</td>
<td>6.00*</td>
<td>M1 &lt; M2</td>
</tr>
<tr>
<td>% (+ )</td>
<td>33.3</td>
<td>61.1</td>
<td>55.6</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Adequacy/Zync n(+)</td>
<td>10</td>
<td>10</td>
<td>12</td>
<td>0.89</td>
<td>SL</td>
</tr>
<tr>
<td>% (+ )</td>
<td>55.6</td>
<td>55.6</td>
<td>66.7</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Adequacy/Iron n(+)</td>
<td>13</td>
<td>15</td>
<td>12</td>
<td>1.40</td>
<td>SL</td>
</tr>
<tr>
<td>% (+ )</td>
<td>72.2</td>
<td>83.3</td>
<td>66.7</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Adequacy/Vitamin C n(+)</td>
<td>8</td>
<td>12</td>
<td>12</td>
<td>3.56</td>
<td>SL</td>
</tr>
<tr>
<td>% (+ )</td>
<td>44.4</td>
<td>66.7</td>
<td>66.7</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* P<0.05
Colonization has not always caused loss of appetite and subsequent weight loss in all cases, especially in those who presented good nutritional status. Generally, since malnutrition was mild, and the patients showed nutritional improvement during follow-up, the lung colonization with *P. aeruginosa* influenced the weight of most patients. Shale confirms the evidence by mentioning that chronic malnutrition increases the risk of lung colonization and affects the immune response.38

The hospitalization of patients was not necessarily correlated to the aggravation of the nutritional status, since in this study most patients were occasionally hospitalized or underwent a scheduled hospitalization to be decontaminated of *P. aeruginosa*, and they did not lose weight either before or during the period of decontamination (Figure 1).

In our study, we demonstrate a significant improvement of height/age after continuous, oral nutritional intervention. There were no significant alterations in the weight/height z score during follow-up. There was an increase in weight/age during follow-up, although it was not significant (Table 2).

A recent longitudinal study carried out in Brazil, with a larger sample size than the one used in this study, revealed a significant increase in the weight/age z score, after 13 and 43 months of continuous, oral nutritional intervention in patients aged between six months and 18 years old. However, the results were similar to the ones in this study for patients younger than five years old, with a significant increase in the height/age z score.59 Luder et al. did not identify a significant improvement of the height z score in patients aged between two and 27 years old, with significant increase in weight after continuous, oral nutritional intervention.40

The study by Gaskin et al. tried to improve the nutritional status through oral nutritional supplementation, but they did not have significant results. However, in the group (4-16 years) that received the nocturnal diet by means of gastrostomy, a significant improvement was verified regarding the nutritional indicators after 18 months of intervention.41 O’Loughlin et al. only observed significant improvement of weight, lean body mass and fat body mass after the intervention by means of a nasoenteric feed tube during six months in patients (7-27 years old) that did not obtain improvement through oral supplementation.42

Other studies that performed aggressive nutritional intervention during a minimal period of 12 months, with nocturnal diet through gastrostomy, demonstrated improvement of the nutritional status. Levy et al. observed significant improvement of height and weight of patients aged between 4.9 and 21.5 years old.5 Other studies mentioned significant increase of weight in patients older than seven years.43,44 Generally, the studies that performed oral supplementation presented difficulties improving the nutritional status of patients, since the aggressive nutritional intervention presents better results from this point of view.

In this study, breastfed infants presented more significant improvement regarding anthropometric indicators and body composition after nutritional intervention, if compared to the other age groups. At the beginning of the follow-up period (M1), this group had the greatest deficit of height and weight. Similarly, other studies confirmed partial or complete height recovery during the first years of life, which may happen faster when there is early nutritional intervention. This fact highlights the importance of an early diagnosis by means of neonatal screening at birth,55-49 with the dosage of immune responsive trypsin in the blood flow.50

We demonstrated significant improvement in the arm muscle area (AMA) after nutritional intervention, although some patients did not reach the 5th percentile of Frisancho’s table, which is considered the minimal rate of adequacy.16 The fat body mass and the percentage of body fat did not present any significant alterations during follow-up (Table 3). Some studies that compared patients to a control group observed that children,51-53 teenagers52 and adults54-56 have lean body mass and fat body mass values below the recommended rates, which reinforces that, even though there is an improvement of the body composition, some patients are not able to reach the normal rates.

With similar results, Greer et al. verified improvement of the body composition of breastfed infants that were diagnosed at the neonatal screening, after continuous nutritional intervention up to 12 months old, and they were prone to having less fat body mass.45 Some studies using nocturnal diet by means of gastrostomy, for a minimal period of 12 months, revealed not only the significant improvement of lean body mass, but also a significant increase in fat body mass.5,43,44

Therefore, it is possible to verify that nutritional recovery varies a lot, that is, while some studies demonstrated improvement of height or lean body mass, other studies demonstrate weight gain and increase in fat body mass, or both. In the present study, there was a deviation of energy intake towards height gain, with increase in the growth rate and in lean body mass. It also happened because patients already presented good weight/height rate from the beginning. We believe that there must be, besides the quantitative factors, other factors related to food intake that contribute to the gain of lean body mass and height. According to Abrams, the antioxidant nutrients may interfere with the pulmonary condition and nutritional status.57

In this study, we verified that most patients reached the energy percentage recommended after nutritional intervention (Table 4). However, with regard to the percentage of appropriate patients, similarly to other studies, we report the difficulty of reaching the recommended rate of 120% of energy and 40% of lipids for all patients50,58-61 (Table 5). Some authors affirm that it is not necessary to reach 120% of the RDAs (Recommended Dietary Allowances) for energy during all phases of the disease.46,62,63 Wooton et al. pointed out that the RDAs are
calculated according to age and not weight, which suggests that energy requirements are overestimated.\textsuperscript{64} It is possible that in our study some patients did not need to reach 120% of energy due to the smaller amount of pancreatic-insufficient subjects (61%) (Figure 1) than the amount referred to in the literature (85%), and also due to the prevalence of patients with mild clinical condition. According to the results of the Shwachman score, patients remained stable during the follow-up period, which shows statistically significant increase of the mean and median between M1 and M3, even though there were no alterations in the score classification. These results demonstrate that the clinical and nutritional treatment were efficient during the follow-up period, since this score was broadly used in order to evaluate the clinical condition in the most important centers for the treatment of cystic fibrosis (Table 1). Through a descriptive comparison, we verified that among five breastfed infants studied, four infants presented the excellent classification of the Shwachman score at the end of the follow-up, with positive alteration of the score. The other age groups did not show the same intensity of alteration.

During the visits between follow-up moments 1, 2 and 3, the nutritional guidance of the patient was reinforced according to inadequate food intake or nutritional deficiencies. Especially, for patients with deficient nutritional status, we recommended the adjustment of the food intake scheme (number of meals, interval between meals, distribution of macronutrients among the meals, hypercaloric substances) and lipidic supplementation with raw soy oil.

Some studies have employed vegetal oils in order to increase the energetic value, the percentage of fat and fat acids that are essential for good nutrition.\textsuperscript{46,65} Thus, we were especially concerned about the increase of the energetic density with soy oil, since it does not change the volume ingested by the patient, it presents low cost, and if compared to other kinds of oil, it is the one that presents the best ratio of Omega-3 and -6 fatty acids.\textsuperscript{66} There was good compliance with the use of pancreatic enzymes and supplementation with soy oil, since the multidisciplinary work provided a good relationship between the team and the patient (Figure 2). The control regarding the compliance with the use of nutritional supplement and enzymes was performed at all nutritional follow-up visits, in the anamnesis carried out with the patient and the guardian.

We detected increase in the mean amount of protein ingested at moments 2 and 3, however it had already surpassed the RDAs at M1, with proportionally adequate percentage (10-15%) regarding the total energy value (Table 4). Similarly, other studies suggest that usually protein expenditure easily surpasses the RDAs for age.\textsuperscript{40,41,46,58}

The amount of patients with appropriate calcium intake almost doubled at M2 and M3, compared to M1. We have not observed significant alterations of mean zinc intake and number of patients with appropriate values of zinc during follow-up. There was a significant increase in iron intake at M2 and M3, compared to M1. However, the number of appropriate patients practically remained unchanged during follow-up. The difficulty reaching the recommended value for zinc and iron might have occurred due to the anorexia phases, during which the most severely affected meals were lunch and dinner, exactly the meals at which there is greater iron and zinc intake. Anemia was detected in eight (44.4%) patients during the follow-up period, three cases of breastfed infants and five cases of preschoolers and school-age children. Both, preschoolers and school-age children, presented mild anemia (Figure 1). There was a tendency to improve vitamin C intake at moments 2 and 3, compared to M1, with remarkable increase of the number of appropriately

![Figure 2](image.png)

**Figure 2** - Patients with cystic fibrosis and compliance to the use of nutritional supplement and enzymatic supplementation
nourished patients after nutritional intervention (Tables 4 and 5).

Kawchak et al. mentioned that it is usually possible to reach the recommended rate of micronutrients, according to the RDAs, for patients with cystic fibrosis. However, studies in this area are rare and they hardly ever mention the percentage of appropriate patients regarding the intake of micronutrients and energy. They usually just present the mean value ingested, which might be appropriate, although they do not represent the reality of individual cases.

The limitations of the study such as sample size, variability of the clinical status and age groups, have not allowed us to accomplish more detailed statistical analysis in order to compare the subgroups. Taking into consideration the reduced number of longitudinal studies about nutritional status, body composition and food intake regarding this pathology, we believe our study should be published, in spite of the limitations that are inherent to our environment.

In short, the nutritional intervention offered to patients with cystic fibrosis showed that the energy supplied through food intake was used mainly in order to increase the growth rate, which was demonstrated through the improvement of the height/age rate and the amount of lean body mass after the intervention. Breastfed infants presented more significant improvement of the nutritional indicators compared to the other age groups. We concluded that breastfed infants had better ability to reach nutritional improvement compared to the other age groups. This is easily understandable if we consider that their lung was less affected and they had avid appetite, which contributed to the efficacy of the clinical and nutritional treatment. The total analysis of patients regarding the weight/height rate and fat body mass was not altered, suggesting that height gain was a priority, compared to weight gain. The compliance with the nutritional supplementation, the correct use of pancreatic enzymes and the improvement of micronutrient intake also contributed to nutritional improvement.

Nutritional education is necessary in order to improve the patient’s prognosis, and it is more efficient when it is performed sequentially and through a good relationship between the professional team and the patient. Control of the nutritional status, associated with the advances in the clinical and physical treatment, early diagnosis and greater efficacy of medication lead to an increase in patients’ life expectancy.

References


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