

# Considerations on homevisits to adults with cystic fibrosis: an exploratory-descriptive study

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## Abstract

*Objective:* Discuss possible benefits of nutritionist home visits as a follow-up strategy of nutrition therapy to adults with cystic fibrosis. *Method:* Descriptive exploratory study with 14 patients aged 18 or older, both males and females, registered in the Adult Cystic Fibrosis Care Program of the Cystic Fibrosis Association of Rio de Janeiro, in Brazil. Data were collected from medical records and through interviews: socioeconomic, demographic, nutritional, and clinical data as well as information on nutritionist home visits and nutrition therapy. *Results:* the research participants reported that nutritionist home visits can benefit their treatment by improving their knowledge about the disease, nutrition and self-care. There are indications that nutritionist home visits can help establish a relationship of trust between nutritionists and CF patients and their families. This is a prerequisite for better results in clinical follow-up. *Conclusions:* nutritionist home visits may be an important strategy for nutrition therapy follow-up with cystic fibrosis patients. Thus, it should be adopted in the Unified Health System as a health care measure for people with special dietary needs, because it can raise further awareness of the disease, assist in self-care, improve patients' nutritional status and, consequently, provide greater control over cystic fibrosis.

**Keywords:** Cystic Fibrosis. Adults. Home Visits. Nutrition Therapy.

## Introduction

Cystic fibrosis (CF), also known as mucoviscidosis, is an autosomal recessive, chronic genetic disease, with systemic manifestations; it is characterized by dysfunction of the exocrine glands, including the pancreas, sweat glands and mucous glands, and it affects the respiratory, digestive and reproductive systems.<sup>1</sup> The disease is associated with mutations in a gene that encodes a membrane protein, the transmembrane conductance regulator in cystic fibrosis, which is a c-AMP-dependent chloride channel which reduces chlorine transport in the apical surface of epithelial cells.<sup>2</sup>

The diagnosis of CF is still difficult to make and little known in Brazil. Immunoreactive trypsin can be measured at birth by the heel-prick screening test. Although it is not considered a conclusive diagnosis, it is an important step in screening.<sup>3</sup> Early diagnosis of CF can promote significant improvement in nutritional status and early intervention in pulmonary disease, thus reducing mortality.

In Brazil, screening for CF was established in 2001 by the Ministry of Health but currently there are few states in the country that perform neonatal screening for CF.<sup>4</sup> The state of Rio de Janeiro started conducting the test as of September 2010.

The gold standard for diagnosing CF is the sweat test, which is the quantitative measurement of chloride in sweat, obtained by the method of pilocarpine iontophoresis.<sup>5</sup> People are referred for this test when they have a family history of CF, positive screening test of the newborn, signs and symptoms suggestive of CF and a suspected active phenotype.

Genetic testing is also used to detect the presence of CF-related mutations.<sup>1</sup> The main symptoms are: salty sweat, recurrent cough with mucus, recurrent pneumonia, weight gain and loss of fat in feces.

In Brazil, it is estimated that the prevalence of the disease is 1: 10,000 live births, although there are variations in the frequency of mutations in different geographical regions. In the southern region, the prevalence is similar to that of the Central European Caucasian population of 1: 2,000 to 1:1,500 live births.

According to 2011 data, in the state of Rio de Janeiro, 21% of the 209 CF patients registered in the Cystic Fibrosis Association of Rio de Janeiro\* (ACAM) were in adults aged 18 years or older.

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\* The mission of the Cystic Fibrosis Association of Rio de Janeiro (ACAM) is to assist people with cystic fibrosis in the State of Rio de Janeiro. Its main objectives are: to support and fight for the rights of CF patients and their families; raise awareness of the disease; stimulate and support studies and research for the treatment and cure of cystic fibrosis. For this purpose, the association has health professionals from various fields, such as nutritionists, physiotherapists, social workers and psychologists.

Given the increasing life expectancy of CF patients, studies with adults are needed in order to seek improvements in health care provision and prognosis for these cases.

A survey, conducted by ACAM in 2007 to assess the profile of CF patients in Rio de Janeiro, found that some patients did not follow the treatment properly, often because they were ill-informed or faced socioeconomic hardship.<sup>6</sup> There was smaller adhesion of CF patients to treatment as regards dietary counseling and physical activity, which required strategies to improve their engagement.<sup>7</sup> In this perspective, the research problem is the lack of adherence strategies to nutritional treatment that can meet this growing segment of the population.

Home visits (HVs) are considered as a strategy that can contribute to adult CF patients' adherence to treatment as adequately as possible, so that they can perform routine and daily functions as normal adults, e.g. work, study, have relationships and enjoy themselves. HVs, in the field of health care, are understood as visits made by a health professional to a patient's home, with the purposes of health care, learning or research.<sup>8</sup> HVs, therefore, should be made by such professionals within an educational perspective, aiming at self-care<sup>9</sup>

Although there is no cure for CF, it can be treated, mainly through prescription drugs, including digestive enzymes, oral and inhaled antibiotics, anti-inflammatory drugs, mucolytic drugs and bronchodilators, as well as by daily chest physiotherapy and maintenance of nutritional status. Nutritional follow-up plays an essential role in the treatment of CF, as it influences the survival and quality of life of these patients.<sup>10</sup> In this sense, nutritionist home visits (NHVs) can help patients better understand the disease, acknowledge the importance of treatment and make adjustments in everyday life so as to achieve a better nutritional status and possibly avoid recurrent complications. The objective of NHVs is to adapt the treatment proposed at the clinic to the home environment, answer patients' questions and provide information about the disease, its nutritional aspects and the possibility of increased survival of CF patients. It is believed that such activities can promote adherence to treatment.

According to the National Food and Nutrition Policy (PNAN), special dietary needs, whether restrictive or supplementary, are those related to individuals with metabolic or physiological changes that cause temporary or permanent changes associated with the biological use of nutrients or (enteral or parenteral) food intake.<sup>11</sup> Examples of special dietary needs are inborn errors of metabolism, celiac disease, HIV/AIDS, food intolerance, food allergies, eating disorders, prematurity, nephropathy, among others.

It should be noted that the documents on Nutritional Health Care of the Unified Health System (SUS) establish that the population's health demands and needs are to be prioritized, by considering the most prevalent and relevant ones and observing risk and vulnerability criteria.

In addition to the priorities indicated in PNAN, such as obesity, chronic diseases, malnutrition and specific nutritional deficiencies, another demand for SUS is nutritionists' attention to special dietary needs.

The fact that SUS has established strategies such as NHVs for promoting the treatment of CF patients, is crucial for meeting the special dietary needs of this particular population.

In particular, home visits (HVs) are a work methodology aimed at education, health care and research, performed by professionals who visit the home of patients, SUS users or recipients of a service.<sup>12</sup> In this context, a preliminary aim of this study is to discuss possible benefits of NHVs as follow-up strategy of nutrition therapy in adult CF patients treated in the state of Rio de Janeiro, Brazil.

## Methodology

This is a descriptive and exploratory study with adult CF patients treated through NHVs, residents of the metropolitan area of the State of Rio de Janeiro, regularly seen at the Adult CF Program of the Piquet Carneiro Polyclinic (PCP) of the State University of Rio de Janeiro and registered in ACAM, from April to August 2011.

The starting point of the study was the analysis of 19 medical records of CF patients. The following inclusion criteria were applied: both male and female patients, aged 18 or older, diagnosed with CF by the presence of at least one of the typical clinical manifestations of the disease and at least two doses of chlorides in sweat by pilocarpine iontophoresis with values above 60 mEq/L, or by the presence of two known CF-related mutations by genetic diagnosis.

Exclusion criteria were the following: patients who were hospitalized during home visits; patients whose home visits could not be scheduled, for whatever reason; and those with severe malnutrition characterized by body mass index (BMI) below 15 kg/m<sup>2</sup>. Thus, 14 adults were considered as subjects of this study.

The survey was conducted in two locations: the first location was the Piquet Carneiro Polyclinic, where preliminary demographic and anthropometric data were collected, such as weight, height and body mass index before and after NHV; the second location was the CF patient's home.

Data collection was performed according to the following steps, described in Table 1: analysis of medical records; verbal approach; collection of data from medical records; scheduling of visits; home visits; application of data collection script at first NHV; follow-up visits; application of form and new data collection from medical records.

**Table 1.** Data collection steps. 2011.

1st STEP	2nd STEP	3rd STEP
<p data-bbox="191 316 506 420">Selection of patients based on inclusion and exclusion criteria.</p> <p data-bbox="234 471 465 538">Analysis of medical records.</p>	<p data-bbox="552 316 863 460">Outpatients were approached verbally on the day of their visit to the health unit.</p> <p data-bbox="548 511 868 655">In case of acceptance, data were collected from their medical records, e.g., weight, height and BMI.</p>	<p data-bbox="909 316 1215 384">Nutritionist Home Visits were scheduled by phone.</p> <p data-bbox="899 434 1225 538">The next data collection step took place between 1-2 weeks.</p>
<p data-bbox="291 720 408 748">4th STEP</p> <p data-bbox="201 797 501 900">First home visits were made and the nutritional form was applied.</p> <p data-bbox="205 988 496 1092">The next data collection step took place within 1 week.</p>	<p data-bbox="648 720 765 748">5th STEP</p> <p data-bbox="555 797 858 900">The other two visits were made and advice was provided.</p> <p data-bbox="555 951 858 1019">The nutritional form was applied.</p> <p data-bbox="548 1070 865 1137">The next step was conducted within 8 weeks.</p>	<p data-bbox="1005 720 1122 748">6th STEP</p> <p data-bbox="899 797 1225 940">Data (weight, height and BMI) were collected from patients' medical records at the Polyclinic.</p>

The profile of patients was identified from data in the form, particularly the variables sex, age, skin color, method and age of diagnosis, schooling, labor market entry, family income, main complaints about diseases, nutritional supplements and prescription drugs. Dietary recalls were also used to assess food intake, because they are a valuable tool to identify patients at nutritional risk and, thus, define the best dietotherapy.

Based on the information completed on the nutritional form, the researchers developed relevant guidelines to suit the nutritional management of patients' environmental and economic conditions. The health professionals at ACAM make regular HVs and, in the case of this research,

HVs were made by nutritionists. It is noteworthy that, at the time of the visit, the presence of the CF patient, their main caregivers and the whole family is crucial.

**Table 2.** Professional procedures during the first NHV and subsequent visits. 2011.

Professional procedures during the NHV.	Explaining Cystic Fibrosis
	Providing explanations about keeping an ideal body weight
	Explaining the nutritional treatment and the importance of adherence to it
	Aligning treatment with patient's daily routine and needs
	Adapting diet to prescription received at the clinic
	Answering questions
Follow-up on subsequent visits	Resuming dietary routine and treatment in general
	Answering new questions
	Checking inhalator
	Approaching new issues such as selection of foods and respective combinations, cleaning, packaging, storage, thawing, use of food and cooking technique to preserve the nutrients
	Readapting dietary routine and treatment when needed

To discuss possible benefits of NHVs as a follow-up strategy of nutrition therapy with CF adults, an analysis was made of the answers to the form applied at the fifth stage of data collection, as shown in Table 1.

The research project was approved by the Research Ethics Committee of the School of Medicine of the University Hospital Antônio Pedro (HUAP) under CAAE protocol: 6099.0.000.258-10, on 17/12/2010, with adherence to the principles governing the conduct of research with human beings, according to Resolution No. 196/96 of the National Health Council. Patients were informed about the purpose, objectives and methodology of the research, and those who agreed to participate signed an informed consent form.

## Results and discussion

Because CF is an autosomal recessive disease, where both parents carry the recessive gene, it affects men and women equally. There is no evidence reporting that sex influences the disease or its progress.

In this research, according to the data collected, males outnumbered females, and the mean age of the females was higher than that of males.

Table 3 shows the results of the analysis of variables “skin color”, “method and age of diagnosis”, “schooling”, “family income” and “labor market entry”.

**Table3.** Skin color, age at diagnosis, method of diagnosis, schooling, family income and labor market entry of research participants. Rio de Janeiro-RJ, 2011.

Variables	N	%
<b>Skin color</b>		
White	5	35.7
Non white	7	50.0
Did not know	2	14.3
<b>Age of diagnosis</b>		
At birth	2	14.3
Between 1 and 21 years	10	71.4
After 21 years	2	14.3
<b>Method of diagnosis</b>		
Sweat test	12	85.7
Two mutations for CF	1	7.1
Neonatal screening	1	7.1

Variables	N	%
<b>Schooling</b>		
Attending an undergraduate program	3	21.4
Technical high school	1	7.1
Secondary School - Completed	6	42.8
Secondary School - Not Completed	3	21.4
Primary School - Not Completed	1	7.1
<b>Family income</b>		
1 - 2 minimum wages	3	21.4
>2 - 5 minimum wages	6	42.8
>5 minimum wages	5	35.7
<b>Labor market entry</b>		
Formal employment	5	35.7
No formal employment	9	64.3
Total	14	100.0

CF is prevalent among Caucasians, especially of European descent; incidence is one out of every two thousand live births, unlike people of African descent, whose indicator is 25 times greater.<sup>13</sup> However, one should take into account the great miscegenation that occurred in Brazil, which results in new features in the profile of the population. This fact is emphasized when people use a large number of expressions to translate the color of their skin - dark, mulatto, light brown, dark brown, brown - in addition to numerous other shades existing between white and black; there are also indigenous Brazilians, and descendants of Japanese, Chinese, Koreans and Arabs, who cannot specify their skin color among the options offered in population surveys.



Mean age of diagnosis found in this study is 8.1 years; it is considerably higher than the one found in studies in northeastern Brazil: 3.9 years of age.<sup>14,15</sup> It was reported that the median age of diagnosis in the US was six months.<sup>16</sup> This research showed that most people were diagnosed for CF by application of the sweat test, confirming the findings of the literature.

Early diagnosis is important for prophylaxis of pulmonary infections, as well as for pancreatic enzyme replacement in people who have pancreatic insufficiency, thereby increasing nutrient absorption, improving their nutritional status, reducing the complications resulting from the disease and increasing survival.<sup>17</sup>

The disease does not cause any impairments to learning, and CF patients may have as much educational development as a healthy person. It is known that higher levels of education can improve patients' nutritional status and adherence to treatment.<sup>18</sup>

Family income can be considered an important element in the analysis of health conditions, because it translates into the possibility of acquisition and use of goods and services that are essential to the maintenance of health, e.g. food, housing, clothing and sanitation,<sup>16</sup> especially in the case of CF patients. In a study with CF children, it was found that risk of death is 3.65 times higher in the poorest patients, who also had worse pulmonary function and nutritional status.<sup>19</sup> It should be noted that there are families in this research whose income is below two minimum wages. This condition may impair the treatment, because of its high costs. The State provides prescription drugs, but there are other expenses involved, for example, supplementary diet, as well as increased household electricity consumption, because of daily use of inhalators and cooling, as required by some medicines.

In its turn, labor market entry may be impaired due to constant hospital admissions; CF patients are often unable to keep up a steady job, because they often depend on sick leave. Thus, these people are disadvantaged in terms of work activities.

Tables 4 and 5 show the main complaints of patients and frequency of use of nutritional supplements.

**Table 4.** Major complaints of CF patients. Rio de Janeiro-RJ, 2011.

Major complaint	N	%
Respiratory fatigue / dyspnea	8	57.1
Abdominal discomfort	3	21.4
Headache	2	14.3
No complaints	1	7.1

**Table 5.** Frequency of use of nutritional supplements. Rio de Janeiro-RJ, 2011.

Nutritional Supplements	Frequency of intake					
	Once a day		Twice a day		3 times a day	
	N	%	N	%	N	%
Powder hypercalorie	3	21.4	3	21.4	-	-
Supplement Liqu	3	21.4	2	14.3	1	7.1
Medium chain triglyceride	1	7.1	1	7.1	-	-

It is known that both physical fatigue and dyspnea may compromise the nutritional status of CF patients,<sup>20</sup> hence the need to use nutritional supplements to increase the daily energy intake for weight gain purposes.

Nutritional intervention in CF adults should begin early, avoiding deterioration in lung function and producing a positive effect on survival. All CF patients should be assessed regularly, and their nutritional status and energy intake should be monitored in order to ensure adequacy. The nutritional recommendation includes a high-fat diet, with 35 to 40% of calories from that source. When compared to a person without the disease, CF patients may require 120-150% of the estimated daily energy requirements. Oral supplements can be used in selected cases. The goal is to maintain body mass index (BMI) between 20 and 25 kg/m<sup>2</sup>; BMI below 19 kg/m<sup>2</sup> indicates significant malnutrition and the need for immediate intervention, which may require hospital admission of the person under these conditions.

The data showed that most subjects used food supplements. The frequency of use ranged from one to three times a day. The patients used high calorie powder and liquid supplements, and medium chain triglyceride in combination with these supplements.

Table 6 shows the number of meals per day of the patients interviewed.

**Table 6.** Number of meals per day eaten by the research participants. Rio de Janeiro-RJ, 2011.

Number of daily meals	Participants in the survey	
	N	%
3	2	14.3
4	4	28.6
5	2	14.3
6	3	21.4
More than 6	3	21.4

The number of meals per day ranged from three to more than six, but the vast majority performed more than five meals per day. During the interviews, the patients also reported the difficulties they faced when adapting the dietary prescriptions, e.g. regular hours, adaptation of certain foods to their socioeconomic conditions and use of other prescription drugs, and their daily study and labor duties.

Prescription drugs used by participants were digestive enzymes, oral and inhaled antibiotics, anti-inflammatory and mucolytic drugs, and bronchodilators. Most of the patients used inhalation devices at least twice a day, and took two oral medications daily. Many patients used pancreatic enzymes to enhance absorption of nutrients, and they also needed fat-soluble vitamins and oral anti-inflammatory drugs. Other drugs were taken according to individual requirements.

All nutritional supplements and drugs used by CF patients were provided on a monthly basis through the Department of Health and Civil Defense of the State of Rio de Janeiro, in two drug-dispensing locations: Fernandes Figueira Institute, which dispenses medication to children and adolescents up to 19 years of age on a monthly basis; and the Piquet Carneiro Polyclinic, which dispenses medication to adults throughout the state.

Table 7 shows some of the impressions of respondents about nutritionist home visits (NHV) in terms of treatment adherence.

**Table 7.** Impressions of research participants about Nutritionist Home Visits (NHVs), Rio de Janeiro-RJ, 2011.

Questions	Yes	No
Have you ever received HVs by any health professional before?	9	5
Do you believe your treatment adherence has improved after NHVs?	9	5
Do you find it hard to gain weight?	10	4
Do you wish to continue the nutritional treatment?	13	1
Can you follow the nutritionist's prescription?	10	4
Do you think treatment is easier to follow with NHVs?	9	5
Do you still have questions about the nutritional treatment?	0	14
Did you feel supported during NHVs?	14	0

In the present research, these are the major statements respondents made during interviews about NHVs in the nutritional treatment of CF:

*Nutritionist home visits helped me organize the times of my meals, and the physical exercises I did during the week also helped me improve in my treatment. [Subject 3].*

*I think it's much better than before. After the visits, I started to eat better and do things that I didn't used to do before. [Subject 4].*

*Yes, because after the visit you feel more supported, as if someone or some people are monitoring you, watching you. You feel useful and end up doing all you were asked more easily. [Subject 6].*

*Yup. The nutritionists explain everything and answer my questions. I felt more confident to start the treatment. [Subject 7].*

*Yes. I learned how to eat better and combine foods right, and I also learned to associate food with exercise. [Subject 9].*

*Yes, they told me about the healthiest foods and said I should eat a greater number of meals per day. [Subject 13].*

According to the above reports, NHVs were considered as beneficial in the nutritional treatment of CF. In order to better assess the possible contribution of NHVs to CF patients, the form included the following question: What have NHVs meant to you?

*"NHVs meant more confidence when going on a nutritional diet. [Subject 1].*

*A great help to continue the treatment at home, and get answers to all my questions about nutrition. [Subject 3].*

*It meant a great improvement to my life. The advice and attention I was given improved my treatment in many respects, but mostly the psychological one (the issue of acceptance), which is very important when you're diagnosed with the disease. [Subject 6].*

*An opportunity to follow the treatment correctly. [Subject 12].*

*It meant improvement in the quality of the food I eat. [Subject 13].*

*It was good because I could understand better how I should follow the treatment. [Subject 14].*

The patients may have felt more supported, more confident after receiving clarification and information, and they recognized the importance of NHVs in their treatment. They also highlighted the emotional support arising from this strategy. Borges argues that patients require a follow-up that involves not only proper therapeutic care but also an approach that can encompass the “human character” of the practice.<sup>8</sup> Thus, the presence of health professionals, whether at home or at the health unit, assisting the patients and establishing bond of trust, is a prerequisite for the clinical follow-up to happen. This bond of trust can foster greater confidence and encourage patients to believe that their treatment is possible. There is a better relationship between health professionals and patients when their encounter takes place in a context that fosters new social relations that are more favorable to treatment.

## Final remarks

Advances in general care directed to CF patients can provide growth in the adult population with this disease and increase the demand for development programs to meet the specific needs of these people. Given that several patients from this study had late diagnosis, policies have to be developed for early diagnosis and treatment as a way to reduce the risk of higher impairments in adulthood.

The results of this study suggest that NHVs can be an important follow-up strategy of nutrition therapy for CF patients. Thus, it should be included in the National Health System as a health care measure for people who have special dietary needs. This can expand their knowledge, assist in self-care, improve their nutritional status and, consequently, provide greater control of cystic fibrosis.

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