Dietary patterns of Brazilian adults with neurofibromatosis type 1

Patrones dietéticos de adultos brasileños con neurofibromatosis tipo 1

ABSTRACT

Neurofibromatosis type 1 (NF1) is an autosomal dominant genetic disease characterized by multisystem involvement such as bone, muscle, endocrine, ophthalmologic, cardiovascular, central and peripheral nervous system, cognitive capacity, voice, and oral motor disorders. Nutritional studies in individuals with NF1 have been performed recently. While a previous study showed an inadequate nutrient intake in patients with NF1, the dietary patterns of this population have not yet been widely studied. This study aimed to characterize dietary patterns in Brazilian adults with NF1. Sixty NF1 individuals (51.7% women), ≥18 years of age underwent nutritional assessment including laboratory analysis, anthropometrics, and eating habits recorded on a food frequency questionnaire. Cluster analysis was used to distinguish between dietary patterns. Hypothesis tests were used to compare data. Two groups with distinct patterns were identified, “Healthy” (46.7%) and “Western” (53.3%). These groups were similar in most of the socioeconomic, anthropometric, demographic and laboratory parameters evaluated. However, the upper-arm total area and upper-arm muscle area (UAMA) were lower in the Western group than those in the Healthy group [59.8 (25.7) cm² versus 65.6 (28.3) cm², P=0.049; 35.6±12.4 cm² versus 43.8±15.0 cm², P=0.024, respectively]. In this study, most individuals with NF1 had a Western dietary pattern and this group showed a lower UAMA, which may indicate a potential contribution, even in part, of diet in the muscle phenotype in this population. This association between diet and muscle in NF1 individuals requires investigation in further studies.

Keywords: Dietary patterns; Food intake; Neurofibromatosis type 1; Nutritional status; Skeletal muscle.

RESUMEN

La neurofibromatosis tipo 1 (NF1) es una enfermedad genética autosómica dominante caracterizada por la afectación multisistémica, alterando los sistemas óseo, muscular, endocrino, oftálmico, cardiovascular, nervioso central y periférico así como las capacidades cognitivas. Un estudio previo señaló una ingesta inadecuada de nutrientes en pacientes con NF1, pero los patrones dietéticos de esta población aún no han sido estudiados ampliamente. El objetivo de este estudio es caracterizar los patrones dietéticos en brasileños con NF1. Sesenta individuos con NF1 (51.7% mujeres) ≥18 años se sometieron a una evaluación nutricional que incluyeron análisis de laboratorio, antropometría y hábitos alimentarios registrados en un cuestionario de frecuencia alimentaria. El análisis de conglomerados se utilizó para distinguir los patrones dietéticos; las pruebas de hipótesis para comparar datos. Se identificaron dos grupos con patrones distintos, denominados Saludables (46.7%) y Occidentales (53.3%). Estos grupos fueron similares en la mayoría de los parámetros socioeconómicos, antropométricos, demográficos y de laboratorio evaluados. Sin embargo, las áreas total braquial (ATB) y muscular braquial (AMB) fueron menores en el...
INTRODUCTION

Neurofibromatoses are a group of three genetic diseases characterized by dermatological alterations and propensity to several neural tumors. The most frequent form is neurofibromatosis type 1 (NF1), with a prevalence of 1:3,000 live births. NF1 is an autosomal dominant disorder resulting from mutations in NF1, which is located on chromosome 17 and characterized by a dysfunction in neurofibromin synthesis, a protein responsible for suppressing tumor growth. The diagnostic criteria of NF1 were defined in 1978 by a National Institutes of Health (NIH) consensus and are based on clinical features such as cafe au lait spots, dermal and plexiform neurofibromas, Lisch nodules, axillary and/or inguinal freckling, and bone dysplasia. However, as neurofibromin is expressed in multiple cell types, there may be multisystem involvement such as bone, muscle, endocrine, ophthalmologic, cardiovascular, central and peripheral nervous system, cognitive capacity, voice, and oral motor disorders.

Recently, studies on the nutrition aspects in NF1 have begun to be performed. Studies prior to 2014 were limited to anthropometric evaluation of this population. The anthropometric characteristics most often found in NF1 individuals include a high prevalence of low weight, short stature, macrocrania, reduced muscle mass, and lower body mass index. Vitamin D deficiency or insufficiency, low bone mineral density, reduced muscle strength, and intestinal constipation have also been described and may be associated with an inadequate diet.

The first study to evaluate nutrient intake in NF1, based on three 24-hour recalls, suggested that this population consumes an inadequate diet with excessive fat and sodium and deficient fiber and micronutrient consumption, especially magnesium, calcium, vitamin D and pyridoxine.

However, the evaluation of dietary patterns is more comprehensive than that of isolated nutrients or foods. Meals are composed of multiple foods, resulting in interaction and synergy between their various components, which can compromise the effects on the body. Isolated analyses may be inappropriate because they disregard the results of these interactions as modifications to the bioavailability of nutrients and the simultaneous effects of components whose properties cannot be attributed to a single factor. Therefore, the World Health Organization (WHO) recommends the use of dietary patterns as an adequate method to indicate possible associations between diet, health and biopsychosocial aspects involved in the eating process.

Thus, due to the scarcity of studies evaluating food consumption among individuals with NF1 and the lack of studies evaluating dietary pattern among individuals with NF1, this study aimed to characterize the dietary profile of this population and its relationship with anthropometric, sociodemographic and health characteristics.

METHODS

Study design and sample

This observational cross-sectional study included all individuals with a confirmed clinical diagnosis of NF1 according to NIH Consensus diagnostic criteria, aged ≥18 years, of both sexes, who were evaluated between September 2012 and September 2013 in a Brazilian Neurofibromatosis Outpatient Reference Center. The exclusion criteria were musculoskeletal limitations in the upper and/or lower limbs, use of medications that could compromise nutritional assessment, cancer, or the presence of acute and chronic diseases that required specific diets or food intake.

The sample size was calculated based on the results of the study by Souza et al considering 54.9% of inadequate micronutrient consumption and a population of 464 adults in a Brazilian Neurofibromatosis Outpatient Reference Center at that time. A power calculation was performed using Epi Info™. To attain 90% power, a minimum of 59 individuals was required.

Data collection and instruments

A trained registered dietitian performed all data collection. This procedure was adopted in order to avoid bias such as discrepancies between evaluators in anthropometric measurement as well as induction and/or value judgments that could compromise the assertiveness of the responses during the application of the food frequency questionnaire (FFQ).

For sample characterization, information on sociodemographic and health was collected, including age, per capita household income, and physical activity. Individuals with NF1 were asked about the periodicity of each food’s consumption in the six months preceding the interview, with the following options: daily, weekly, monthly, or never consumed. Data on food intake were obtained by means of a semi-quantitative FFQ containing 67 food items. The FFQ was developed and validated by Ribeiro and Cardoso by adapting of a validated questionnaire applied to the Japanese-Brazilian...
community\textsuperscript{18}, excluding foods of Japanese origin. This FFQ was validated for nutritional assessment and actions to prevent non-communicable chronic disease\textsuperscript{17}. Items commonly consumed in the county (cheese bread and cassava starch snacks) were included in our study. As our objective was to qualitatively evaluate dietary patterns of individuals with NF1, only results related to habitual food frequencies of consumption have been presented.

Physical activity level was evaluated using the validated International Physical Activity Questionnaire (IPAQ) short version. Physical activity levels were classified as very light, light, moderate or heavy according to the IPAQ\textsuperscript{19}.

Laboratory test results were also included in this study. Blood levels of glycemia, total cholesterol and fractions, and triglycerides were measured at the hospital's clinical analysis laboratory after the individuals had fasted for 10–12 hours.

For anthropometry data, weight, height, waist circumference (WC), upper-arm circumference (UAC), and triceps skinfold thickness (TSF) measurements followed the protocol described by the WHO\textsuperscript{20}.

Weight was measured using a mechanical scale (Welmy\textsuperscript{6}) with sensitivity of 100g which was checked over and manually calibrated before each weighing. Height was measured with the vertical stadiometer of the scale (Welmy\textsuperscript{6}). Weight and height were used to calculate body mass index (BMI). BMI categories included normal (BMI between 18.5 and 24.9 kg/m\textsuperscript{2}), low (BMI <18.5 kg/m\textsuperscript{2}), and overweight (BMI ≥ 25.0 kg/m\textsuperscript{2})\textsuperscript{20}.

WC was measured at the midpoint between the iliac crest and the last rib. According to the WHO cutoff points, the maximum normal values for WC were 94 cm for men and 80 cm for women\textsuperscript{21}.

UAC was measured using graduated tape at the midpoint between the acromion and the olecranon on the right side of all patients. Tricep skinfold (TSF) was measured at the same location marked for UAC, using a Cescorf\textsuperscript{20} adipometer positioned perpendicular to the fold, with arms relaxed and extended along the body. The folds were measured vertically, 1 cm from the evaluator’s left thumb and forefinger. Measurements were performed in triplicate and means were considered\textsuperscript{20}.

TSF and UAC were used to calculate total upper-arm total area (UATA), upper-arm muscle area (UAMA), and upper-arm fat area (UAFA), using the following equations, according to Frisancho\textsuperscript{22} (π ≈ 3.14):

\[
\text{UATA (cm}^2\text{)} = \frac{(\text{UAC})^2}{(4 \times \pi)}
\]
\[
\text{UAMA (cm}^2\text{)} = \frac{((\text{UAC} - \text{TSF} \times \pi)^2}{(4 \times \pi)} - 10 \to \text{Men}
\]
\[
\text{UAMA (cm}^2\text{)} = \frac{((\text{UAC} - \text{TSF} \times \pi)^2}{(4 \times \pi)} - 6.5 \to \text{Women}
\]
\[
\text{UAFA (cm}^2\text{)} = \text{UATA} - \text{UAMA}
\]

Statistical Analyses

Data were recorded in a double typing process and later compared to evaluate consistency. All statistical analyses were performed using the STATA Version 14.0 software (STATA; Corp, College Station, Texas).

The food items on the FFQ were grouped into 23 categories according to the similarities of nutritional content and culinary use. The frequencies of consumption of these foods were transformed into a daily consumption score. Dietary patterns were identified via cluster analysis and the variables were initially standardized (with mean zero and standard deviation one). The squared Euclidean distance was adopted as a criterion for dissimilarity; the Ward method was applied for conglomerate formation and the determination of group numbers was performed a posteriori using the Pseudo T2 statistic\textsuperscript{23}. For interpreting this analysis, the mean of the standardized scores was used; in other words, frequencies of consumption below or above the average were considered negative and positive factors, respectively. In our study, two dietary patterns were identified, Healthy and Western, considering the foods most consumed by individuals in each group.

Grouped comparisons of qualitative variables were performed using Fisher’s chi-square or exact tests. Qualitative variables were described as absolute and relative frequencies (percentage). Shapiro–Wilk tests were used for continuous variables to analyze their normality and to determine the appropriate statistical test. Normally distributed quantitative variables were described as means and standard deviation (SD). The groups were compared through independent-samples t-tests. Quantitative variables that were not normally distributed were presented as medians and interquartile range (IQR) and were compared by Mann–Whitney U tests. Multiple linear regression analyses were performed to adjust the variables that showed a statistically significant difference between the groups to the confounding factors. P-values <0.05 were considered statistically significant.

Ethical Statement

This study was conducted according to the guidelines laid down in the Declaration of Helsinki and all procedures involving human subjects were approved by the Ethics Committee of the Federal University of Minas Gerais (#81497). Written informed consent was obtained from all subjects.

RESULTS

This study included 60 individuals with NF1 aged 18-64 years (51.7% women). Two groups with different dietary patterns were identified: Western and Healthy. Figure 1 shows the comparisons of consumption between the two patterns based on standardized mean scores from the cluster analyses.

The first pattern, termed Western, contained 32 individuals (53.3%) and was characterized by a frequency of consumption above the average of margarine and mayonnaise, pastry and fried foods, sausages, artificial and alcoholic beverages. The second pattern, termed Healthy,
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included 28 individuals (46.7%) and was characterized by an increased frequency of consumption compared to the average of fruits, vegetables, legumes, nuts, natural drinks (coffee, tea, and fresh fruit juices), white meat, and seafood. Table 1 presents the food distributions for each dietary pattern based on cluster analysis.

Table 2 shows the demographic and socioeconomic characteristics of this study and the comparisons between the dietary patterns. There were no statistically significant differences in age, sex, per capita income, and education level between groups.

No differences were observed in relation to the laboratory test data and the results were all within the laboratory reference range of values (Table 3). Regarding physical activity, individuals were mostly sedentary, forty-seven (78.3%) did not perform regular physical activity or performed only light daily activities, with no differences between groups regarding physical activity level (P= 0.753) (Table 4).

Regarding BMI categorization, the Western pattern included 19 (59.4%) individuals classified as normal weight, while eight (25.0%) were overweight and five (15.6%) were underweight. The Healthy pattern had 16 (57.1%) with normal weight, 11 (39.3%) overweight and one (3.6%) underweight person. There was no statistically significant difference between the groups (P= 0.208) (Table 4).

Both groups were similar in relation to anthropometric parameters, except for UATA and UAMA (Table 4). The UATA was significantly higher in the Healthy group compared to the Western group (P= 0.049). The UAMA was also higher in the Healthy group compared to the Western group (P= 0.024). The UAFA did not differ significantly between groups (P= 0.264) (Table 4). Multiple linear regression analyses were performed to test whether there was any difference in the UAMA between the dietary patterns, after adjusting for sex, age, BMI and physical activity level, with the Healthy pattern considered the referent group. The association between dietary pattern and UAMA remained significant even after adjustments. Adherence to the Western dietary pattern reduced UAMA by 4 cm² regardless of the factors mentioned above (P= 0.018) [CI 95% -7286035 – -.71864] (Table 5).

Figure 1: Comparison of consumption between Western and Healthy patterns based on cluster analyses.
**DISCUSSION**

The results of this study demonstrated two different dietary patterns, Healthy and Western, among individuals with NF1. Individuals with the Western pattern had a lower UAMA than the Healthy pattern group, suggesting a potential contribution of this dietary pattern on muscular aspects of NF1.

In general, balanced diets that are based on fresh and minimally processed foods, mainly fruit and vegetables, are considered Healthy patterns, which correspond to the recommendations of the Dietary Guidelines for the Brazilian Population\(^{24}\). In contrast, the Western pattern was based on energy-dense, low-nutrient-density foods, rich in ultra-processed foods and artificial beverages, and had reduced consumption of fresh foods such as fruit and vegetables. The food consumption by the Western group in our study is similar to that of the dietary pattern of the general Brazilian population\(^{24,25,26}\).

Consumption of a Western Diet can contribute to the low fiber and micronutrient content, as well as to the excessive consumption of fat and sugars and high-energy density foods\(^{27}\). Although this study did not evaluate

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**Table 1.** Food distributions for each dietary pattern in individuals with NF1 based on cluster analysis.

| FOOD                             | Pattern 1 Western (n= 32) | Pattern 2 Healthy (n= 28) |
|----------------------------------|---------------------------|---------------------------|
| Mean                             | SD                        | Mean                      | SD                        |
| Dairy products                   | -0.054 (1.008)            | 0.061 (1.006)             |
| White meats and seafood          | -0.363 (0.554)            | 0.415 (1.224)             |
| Low-fat red meat                 | 0.407 (1.120)             | -0.465 (0.570)            |
| High-fat red meat                | -0.224 (0.762)            | 0.256 (1.179)             |
| Viscera                          | -0.115 (0.643)            | 0.131 (1.295)             |
| Sausages                         | 0.157 (1.158)             | -0.179 (0.764)            |
| Eggs                             | 0.049 (1.259)             | -0.056 (0.600)            |
| Breads and breakfast cereals     | 0.152 (1.126)             | -0.173 (0.820)            |
| Pasta                            | -0.127 (0.834)            | 0.145 (1.160)             |
| Tubers and cereals               | 0.146 (1.171)             | -0.166 (0.746)            |
| Fruit                            | -0.435 (0.389)            | 0.498 (1.237)             |
| Fresh fruit juices               | -0.367 (0.511)            | 0.419 (1.242)             |
| Legumes                          | -0.421 (0.988)            | 0.481 (0.784)             |
| Leafy vegetables                 | -0.289 (0.783)            | 0.330 (1.127)             |
| Non-leafy vegetables             | -0.364 (0.689)            | 0.416 (1.141)             |
| Nuts                             | -0.343 (0.200)            | 0.392 (1.356)             |
| Margarines and mayonnaise        | 0.314 (1.035)             | -0.358 (0.839)            |
| Fats of animal origin            | -0.178 (0.954)            | 0.203 (1.030)             |
| Pastry and fried foods           | 0.253 (1.116)             | -0.290 (0.769)            |
| Sweets                           | -0.023 (0.875)            | 0.026 (1.142)             |
| Artificial beverages             | 0.215 (1.020)             | -0.246 (0.935)            |
| Coffee and tea                   | -0.154 (0.887)            | 0.176 (1.105)             |
| Alcoholic beverages              | 0.012 (1.103)             | -0.014 (0.888)            |

Note: Values are expressed as the means and standard deviation (SD) of the standardized frequency scores of the food groups’ consumption. Positive factors with consumption frequencies higher than the general average are bolded (cluster analysis).
Table 2. Demographic and socioeconomic parameters in NF1 individuals comparing Western and Healthy dietary patterns.

| PARAMETERS                                | All (n= 60) | Western (n= 32) | Healthy (n= 28) | P-value |
|-------------------------------------------|-------------|----------------|----------------|---------|
| Age (y), median (IQR)                     | 34.0 (13.0) | 30.5 (10.0)    | 35.0 (13.5)    | 0.070'  |
| Sex, n (%)                                |             |                |                |         |
| Male                                      | 29 (48.3)   | 14 (43.8)      | 15 (53.6)      |         |
| Female                                    | 31 (51.7)   | 18 (56.2)      | 13 (46.4)      |         |
| Education level, n (%)                    |             |                |                | 0.650'  |
| No education or incomplete primary school | 10 (16.6)   | 4 (12.5)       | 6 (21.4)       |         |
| Primary school                            | 7 (11.7)    | 4 (12.5)       | 3 (10.7)       |         |
| High school or above                      | 43 (71.7)   | 24 (75.0)      | 19 (67.9)      |         |
| Per capita income, n (%)                  |             |                |                | 0.176'  |
| <= 1 BMW                                  | 29 (48.3)   | 14 (43.8)      | 15 (53.6)      |         |
| 1–3 BMW                                   | 29 (48.3)   | 18 (56.2)      | 11 (39.3)      |         |
| > 3 BMW                                   | 2 (3.4)     | 0              | 2 (7.1)        |         |

Note: BMW: Brazilian minimum wage (considering U$175.00 or R$678.00 in 2013); IQR: interquartile range; *: Mann–Whitney U test; †: Fisher’s exact test and chi-square test.

Table 3. Laboratory tests of individuals with NF1 comparing Western and Healthy dietary patterns.

| PARAMETERS                                | All (n= 54) | Western (n= 28) | Healthy (n= 26) | P-value |
|-------------------------------------------|-------------|----------------|----------------|---------|
| Fasting blood glycaemia, median (IQR)     | 82.0 (9.0)  | 82.0 (8.5)     | 82.5 (12.0)    | 0.211*  |
| Total cholesterol, mean (SD)              | 189.3 (6.4) | 185.2 (9.6)    | 193.7 (8.6)    | 0.518†  |
| LDL, median (IQR)                         | 103.3 (58.0)| 99.4 (47.7)    | 116.9 (57.0)   | 0.299*  |
| HDL, median (IQR)                         | 53.0 (20.0) | 52.5 (25.0)    | 53.0 (21.0)    | 0.521*  |
| Triglycerides, median (IQR)               | 77.0 (44.0) | 64.5 (34.5)    | 84.0 (51.0)    | 0.065*  |

Note: HDL: high-density lipoprotein; IQR: interquartile range; LDL: low-density lipoprotein; SD: standard deviation; *: Mann–Whitney U test; †: Student’s independent-sample t-test.

quantiﬁed data, our results are consistent with those of a study of nutrient intake performed previously by our research group, in which NF1 individuals had an unhealthy diet characterized by excessive saturated fat and a deﬁciency of vitamins, minerals and ﬁber. An unhealthy diet might affect the severity of the clinical manifestations of some comorbidities among NF1 individuals, such as constipation, bone alterations, and reduced muscle mass. An inadequate diet may also predispose individuals to increased risks of chronic diseases such as cancer and cardiovascular disease, whose prevalence is high in NF1 individuals.

Currently, there is no effective therapy for the growth
Table 4. Anthropometric and health parameters in NF1 individuals with Western or Healthy dietary patterns.

| PARAMETERS               | All (n= 60) | Western (n= 32) | Healthy (n= 28) | P-value |
|-------------------------|-------------|-----------------|-----------------|---------|
| Weight (kg), median (IQR) | 59.0 (20.5) | 58.5 (21.0) | 59.2 (16.4) | 0.432* |
| Height (m), mean (SD)   | 1.62 (0.10) | 1.64 (0.11) | 1.61 (0.09) | 0.281† |
| BMI (kg/m²), median (IQR) | 22.9 (5.7) | 22.0 (5.3) | 23.8 (5.8) | 0.100* |
| WC (cm), median (IQR)   | 79.2 (21.3) | 73.2 (19.0) | 80.4 (17.9) | 0.118* |
| UAC (cm), mean (SD)     | 28.3 (4.5)  | 27.4 (4.6)  | 29.4 (4.1)  | 0.094† |
| TSF (mm), median (IQR)  | 12.0 (9.0)  | 11.5 (12.5) | 12.0 (6.5)  | 0.888* |
| UATA (cm²), median (IQR) | 61.5 (29.7) | 59.8 (25.7) | 65.6 (28.3) | 0.049* |
| UAFA (cm²), median (IQR) | 23.4 (10.2) | 23.0 (14.2) | 24.7 (7.0)  | 0.266* |
| UAMA (cm²), mean (SD)   | 39.4 (14.2) | 35.6 (12.4) | 43.8 (15.0) | 0.024† |

BMI category – n (%)  
- Normal weight 35 (58.3) 19 (59.4) 16 (57.1) 0.208‡
- Overweight 19 (31.7) 8 (25.0) 11 (39.3)
- Low weight 6 (10.0) 5 (15.6) 1 (3.6)

WC category – n (%)  
- Normal 43 (71.7) 23 (71.9) 20 (71.4) 0.353‡
- High risk 12 (20.0) 5 (15.6) 7 (25.0)
- Very high risk 5 (8.3) 4 (12.5) 1 (3.6)

Physical activity level – n (%)  
- Very light 47 (78.3) 24 (75.0) 23 (82.1) 0.753‡
- Mild to moderate 7 (11.7) 4 (12.5) 3 (10.7)
- Active to very active 6 (10.0) 4 (12.5) 2 (7.2)

Note: BMI: body mass index; IQR: interquartile range; SD: standard deviation; UAC: upper-arm circumference; UAFA: upper-arm fat area; UAMA: upper-arm muscle area; UATA: upper-arm total area; TSF: triceps skinfold; WC: waist circumference; †: Student’s independent-sample t-test; *: Mann–Whitney U test; ‡: chi-square test.

Table 5. Factors related to UAMA, mutiple linear regression adjusting for confounding factors.

| UAMA (cm²) | Coefficient | P>|t| | [95% C.I.] |
|------------|-------------|-------|-----------------|
| Dietary Pattern (ref= Healthy) | -4.002.338 | 0.018 | -7286035 | -.71864 |
| Sex        | 1.150.208   | <0.001 | 827.512 | 1.472.904 |
| Age*       | .0078604    | 0.921 | -.1500877 | .1658085 |
| BMI*       | 2.084.817   | <0.001 | 1.740.716 | 2.428.918 |
| Physical Activity Level | -6.979.846 | 0.223 | -18.32855 | 4.368.862 |

*The categories are shown in table 4. Note: BMI: body mass index; CI: confidence interval; UAMA: upper-arm muscle area.
of neurofibromas and, perhaps, diet can contribute to the control of clinical characteristics of NF1, which needs to be studied in future studies. For example, the study of Esposito et al (2010) conducted with a small and uncontrolled sample, reported that adherence to a curcumin-enriched Mediterranean diet in individuals with NF1 was associated with improvement in nutritional and metabolic parameters and reduction in the number and volume of cutaneous neurofibromas.

In our study, individuals with the Healthy pattern had higher UAMA when compared to that in the Western pattern group. There were no differences in age and physical activity level between the groups. The NF1 population usually presents musculoskeletal disorders, including reduced cross-sectional muscle area, lower muscle strength, and poor motor coordination. Reduced muscle mass and strength may be present since childhood and can compromise the quality of life of this population. Studies in individuals without NF1, especially in the elderly, have shown that healthy eating patterns can positively influence muscle mass. Healthy eating patterns are positively associated with lower limb strength and inversely associated with sarcopenia. In contrast, the high-fat Western Diet may impair muscle metabolism, as it produces increased levels of inflammatory cytokines such as interleukin-1 (IL-1) and decreased levels of insulin-like growth factor-1 (IGF-1), leading to subsequent muscle damage, which may produce muscle hypotrophy and reduced strength. The association between diet and muscle in NF1 requires further investigation.

Individuals with NF1 often have cardiovascular disease, which causes about 16–18% of mortality. Adherence to a Western diet pattern increases the risk of cardiovascular disease compared to that in other patterns, likely due to its association with increased inflammation and cardiovascular disease progression. Inflammation is also associated with the number of neurofibromas in NF1 patients, although the impact of diet on inflammation in NF1 has not been investigated. Despite approaching the recommendations of the Dietary Guidelines for the Brazilian Population, the Healthy pattern in our study showed consumption of some dietary sources of saturated fats, usually classified as a risk factor for cardiovascular diseases. However, it is important to note that our data indicate only consumption; therefore, it is not possible to determine quantities of foods consumed.

Individuals with NF1 are at higher risk of cancer development compared to the general population. Malignant peripheral nerve sheath tumors are the main cause of death in this population. The Western dietary pattern has been linked to an increased risk of cancer, increased obesity, oxidative damage, and reduced antioxidant defense.

The Western pattern is likely influenced by the local food environment, since the Brazilian population went through a process of replacing the consumption of basic and traditional foods with ultra-processed foods between 1974 and 2003, which resulted in increased consumptions of total and saturated fats, maintenance of the excessive intake of sugar, and deficient fruit and vegetable consumption.

In addition, there is evidence that income and education level can influence dietary patterns and food purchases, in which higher income and education favor healthier food choices. In our study, income level and education were similar between groups.

Adherence to the Healthy pattern, observed in 46.7% of the sample, contrasted with the pattern observed in the general Brazilian population and may be related to uncertainty about disease progression and absence of a cure. Until now, there is no scientific evidence for any effect of diet on NF1. There is also no specific treatment that prevents the growth of neurofibromas. Thus, some individuals with NF1 may have food choice concerns, leading them to seek a healthy diet in an attempt to reduce the risk of developing comorbidities or to induce neurofibromas reduction or even attenuate their growth. It is important to report that none of the individuals in our study had prior nutritional monitoring.

This study had limitations regarding the sample and the methods. Non-probabilistic sampling was performed, given the specificity of the population and the disease, and there was no control group. The FFQ method also has limits due to the improper grouping of foods, an incomplete food list, and the reliance on individual memory for the self-reported frequencies. However, to avoid this kind of bias, a validated questionnaire was used and the interviewer was trained to apply this tool. In addition, UAMA is not the best method for assessing muscle mass; future studies should use gold standard parameters to investigate muscle mass in individuals with NF1. Despite these limitations, this study demonstrated differences in the dietary patterns of individuals with NF1, which indicates that nutritional interventions may be an option to improve the quality of life of this population.

In conclusion, most individuals with NF1 in this study had a Western dietary pattern of food consumption. Our results also demonstrated a relationship between the Healthy dietary pattern and greater upper-arm muscle area, suggesting a nutritional potential contribution, in part, to the muscle impairment commonly described in NF1. Further studies should investigate the mechanisms of action of this association as well as the impact of nutrition and diet on clinical characteristics in individuals with NF1. The high prevalence of individuals in the Western pattern group reinforces the importance of nutritional interventions in this population.

**Conflict of Interest:** The authors declare no conflicts of interest.

**Funding:** This work was supported by CAPES; National Council of Technological and Scientific Development-CNPq (#471725/2013-7); FAPESP, (#APQ-00928-11; #PPM-00120-14). The funding sources played no role in the study design and analysis or in the writing of the manuscript or decision to publish.
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