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Body composition in Spinocerebellar ataxia type 3 and 10 patients: Comparative study with control group

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Abstract

Background: Spinocerebellar ataxias (SCAs) are a group of neurodegenerative genetic diseases characterized by movement disorders that can affect nutritional status and body composition. This study sought to assess body composition in SCA3 and SCA10 patients.

Methods: Anthropometric assessments and bioelectric impedance analysis were performed in 46 SCA3 and SCA10 patients and 76 controls of both genders.

Results: Of the patients, 69.6% had SCA3 and 58.7% were women. SCA3 patients had significantly lower percentages of body fat (%BF) than controls (15.0 ± 6.1 vs. 20.6 ± 7.1 ; $p=0.014$) and (22.4 ± 6.9 vs. 30.1 ± 6.0 ; $p<0.001$), respectively. Among the women, there was a statistically significant difference in %BF between SCA3 and SCA10 patients (22.4 ± 6.9 vs. 32.4 ± 4.9 ; $p<0.001$). Male and female SCA3 patients had significantly lower fat-free mass (FFM) than controls [50.6 kg (46.9 – 54.7) vs. 58.6 kg (52.6 – 63.9); $p=0.001$] and [38.2 kg (35.1 – 42.6) vs. 42.8 kg (39.7 – 46.1); $p=0.004$], respectively. Male SCA10 patients also had lower FFM than controls [51.2 kg (47.1 – 55.4) vs. (52.6 – 63.9); $p=0.008$]. Female SCA10 patients had significantly higher FFM than controls and SCA3 patients [45.0 kg (43.3 – 45.6) vs. 42.8 kg (39.7 – 46.1); $p=0.004$] and [45.0 kg (43.3 – 45.6) vs. 38.2 kg (35.1 – 42.6); $p=0.004$], respectively. There was moderate correlation (-0.42) between disease duration and muscle mass (MM), and weak (-0.38) between

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SARA (Scale for the Assessment and Rating of Ataxia) and MM in SCA3. In SCA10, there was no significant correlation between these variables.

Conclusion: Female SCA3 patients had more body composition changes than female SCA10 patients, mainly in relation to FFM. SCA3 and SCA10 patients need nutritional follow-up to minimize body compartment changes.

Keywords

Spinocerebellar ataxias; Neurodegenerative disease; Genetic disease; Nutritional status; Body composition; Anthropometry; Bioelectric impedance; Fat free mass; Muscle mass

Introduction

Spinocerebellar ataxias (SCAs) are a group of neurodegenerative genetic disorders with heterogeneous genotypes and phenotypes.^{1,2}

To date, more than 40 types of SCA have been reported.²

AEC3 is clinically characterized by gait ataxia, extrapyramidal signs, including dystonia, stiffness, and/or bradykinesia, motor neuron disease with fasciculations and myotrophies, weight loss, speech disarticulation, sleep disturbances, and eyelid retraction.^{2,3}

In SCA10, the main phenotype observed is pure cerebellar ataxia, unlike the typical phenotype, which consists of cerebellar ataxia and epilepsy.²

Dysphagia may be present, particularly in advanced stages of the disease,⁴ and patients may also be present with reduced muscle tone and strength.^{5,6} Excessive muscle co-contraction is common in movement disorders and leads to increased metabolic rate.⁷ In addition, a combination of dysphagia and hypermetabolism, as in SCA3, or a sedentary lifestyle can adversely affect nutritional status and body composition.

SCA 3 is the most common form of the disease worldwide, accounting for about 44% $\pm$ SD 25, ranging from 1% to 90%, depending on geographical origin.⁸ Likewise, it is the most frequent mutation in Brazil, identified in approximately 73.5% of the cases, followed by SCA10 with 12% of cases in the southern region. SCA10 is a very rare form of the disease and affects Amerindian populations of Latin America, being described only in Brazil and Mexico.⁹

A knowledge of body composition is essential to determine suitable nutritional therapy and draw up preventive strategies that may have a direct impact on the patient's physical condition and gait speed.¹⁰ Various methods have been developed for measuring body composition accurately 'in vivo,' such as hydrodensitometry, bioelectrical impedance analysis (BIA), isotopic dilution techniques, and dual-energy X-ray absorptiometry. Among these, BIA is the most simple and accessible for use in clinical practice.¹¹

Phase angle (PA) is a parameter derived from the relation between the direct measures of resistance and reactance obtained by the BIA. It is interpreted as an indicator of membrane

integrity and water distribution between the intra and extracellular spaces. PA has also been considered an excellent predictor of body mass and, consequently, of morbidity and mortality in chronic diseases.¹²

To the authors' knowledge, there are no studies on body composition in SCA patients in the literature.

This study therefore sought to investigate the anthropometric characteristics and body composition of two groups of SCA patients (one with SCA3 and the other with SCA10) and compare them between the two groups and with a control group.

Materials and methods

The study was carried out at the Movement Disorders Outpatient Unit, between June 2011 and June 2013 and was approved by the Committee for Medical Ethics in Scientific Research. Patients, or those accompanying them, and controls signed a voluntary informed-consent form after being told about the study aims and protocol.

Subjects

Male and female patients over 18 years old with a clinical and genetic diagnosis of SCA3 or SCA10 were included in the study regardless of their ethnic origin. Exclusion criteria were: presenting with other types of SCA or other causes of cerebellar ataxia such as secondary, recessive, mitochondrial, congenital, or non-hereditary degenerative ataxia; having any type of amputation or metal prosthesis; having some type of hypercatabolic disease such as cancer, AIDS, diabetes mellitus, chronic heart failure, chronic obstructive pulmonary disease, renal and liver failure, hypo- or hyperthyroidism¹²; being wheelchair bound; using a pacemaker; being pregnant or breastfeeding and performing intense physical activity. In this case, intense physical activity, conducted less than 8 hours prior to the examination, includes exercise that increases cardiac output and vascular perfusion and subsequently increases blood flow to skeletal muscle, which warms the muscle and decreases muscle resistance which results in reduced impedance. In addition, intensive activity causes vasodilatation, an increase in skin temperature, which also reduces measured impedance.¹³

All patients were evaluated previously by a neurologist using a standardized protocol to diagnose SCA and determine SARA (Scale for the Assessment and Rating of Ataxia) scores. The SARA scale uses a cumulative score ranging from 0 (no ataxia) to 40 (most severe ataxia) to measure patients' functional and motor skills.¹⁴ Most patients were not taking drugs that could interfere with the emotional state, such as antidepressant drugs. The control group was formed by apparently healthy adult volunteers who had a direct link to the hospital or to a staff member. They were matched according to the physical characteristics of the sample studied according to sex, age (± 5 years) and ethnicity. They were personally invited to participate in the study.

Anthropometry

Anthropometric assessments of all the individuals were performed by the same assessor. Each patient's weight was measured with a MP 4200 digital balance (Balanças Canadá,

Curitiba, Brazil) with a maximum load of 300 kg, four load cells, and a hydraulic system to raise the bed. The subject lay in the supine position with his/her arms and legs extended and relaxed. The patient's height (H) was measured with a wall-mounted stadiometer with an accuracy of 0.1 cm following Gordon et al.¹⁵ If the patient had difficulty standing up, recumbent height was measured.¹⁵ Body mass index (BMI) was calculated using the patient's current weight and height in the equation $BMI = \text{weight in kg}/(\text{height in m})^2$ and the results were compared with the WHO reference standard.¹⁶

Bioelectric impedance

BIA was performed with a BIA 310 bioimpedance analyzer (Biodynamics Corporation, Seattle, WA, USA) following the European Society for Clinical Nutrition and Metabolism (ESPEN) guidelines.¹⁷ To avoid inaccurate bioimpedance measurements, patients were instructed to fast (for 4 hours before the test), not to do any physical exercise (for 12 hours before the test), not to consume alcohol (for 48 hours before the test), not to use diuretics (for 7 days before the test), and to empty their bladder (immediately before the test). During the test, the ambient temperature was maintained at 25°C.¹⁷ Fat-free mass (FFM) was calculated using the single-prediction equation proposed by Kyle et al.¹⁸:

$$FFM = -4.104 + 0.518H^2/R + 0.231W + 0.130Xc + 4.229 \text{ gender},$$

where FFM = fat-free mass (kg); H = height (cm); R = resistance; W = weight; Xc = reactance; and gender = 0 for female and 1 for male.

Body fat was calculated by subtracting FFM from total body mass and was expressed as a percentage by dividing it by body weight and multiplying by 100.¹⁹

PA was calculated directly by the following equation²⁰:

$$PA = \arctan(\text{reactance/resistance}) \times 180^\circ/\pi,$$

Analysis

The statistical analysis was performed with SPSS 22.0 for Mac (SPSS, Chicago, IL). The data were tested for normality with the Shapiro–Wilk test. Parametric variables were expressed as mean and standard deviation ($\bar{x} \pm SD$), and nonparametric variables as median and first and third quartiles. The three groups (control, SCA3, and SCA10) were compared together with one-way analysis of variance (ANOVA) for parametric data and the Kruskal–Wallis test for nonparametric data. Student's t test for independent samples or the nonparametric Mann–Whitney test was used to compare the groups in pairs. The correlation between SARA variables, disease time, and lean mass was analyzed by estimating Pearson or Spearman correlation coefficients, depending on the normality of the variable. A significance level of $p < 0.05$ was used.

Results

Forty-six patients with a clinical and genetic diagnosis of SCA3 (69.6%) and SCA10 (30.4%) were assessed. Due to technical difficulties, two wheelchair patients were not included. The demographic and clinical data are shown in Table 1. Of the 32 SCA3 patients and 14 SCA10 patients, 59.4 and 57.1%, respectively, were female. Mean age was 46.1 ± 9.5 years, and the majority of the subjects were Caucasian. Median SARA score was 12.0, being 36.0 the maximum value, and mean disease duration was 11.4 ± 0.7 years. The male group with SCA10 presented twice as long in comparison to men with SCA3. The control group consisted of 76 healthy volunteers (53.9% female).

The anthropometric characteristics and body composition of the SCA patients and controls of both genders are shown in Table 2.

Among the men, there was a statistically significant difference in height, %BF, and FFM between the two SCA groups and the controls. SCA3 patients had lower values of %BF and FFM than the controls, while SCA10 patients had lower heights and FFM values. Comparison of the two ataxia groups revealed similar results (Table 2).

In contrast, among the women, there was a statistically significant difference in all the variables except PA between the three groups (Table 2). Nearly all the variables were lower for female SCA3 patients than for the control group. In these patients, the estimates corresponding to the body compartments, such as %BF and FFM, were lower than in the other two groups. With the exception of height and PA, the results for SCA10 patients were higher than for the controls. The only variable that was not statistically different between the female SCA3 and SCA10 patients was height (Table 2).

Pairwise comparison of the groups (SCA3 with control, SCA10 with control, and SCA3 with SCA10) for the variables that were statistically significant (*p*-values) in the previous analysis allowed the body changes associated with SCA type to be identified, for males and females (Table 3).

Univariate analysis showed moderate correlations (-0.42) between disease duration and MM, and weak (-0.38) between SARA and MM in patients with ACS3. SARA and duration of the disease showed a moderate positive correlation (0.59). In SCA10, there was no significant correlation between these variables (Table 4).

Discussion

Our results show that body composition changes in SCA patients depend on disease subtype and gender. Generally, SCA3 patients had the lowest values for most of the parameters assessed and SCA10 patients the highest.

The mean body compositions observed in this study show that severely impaired nutritional status is not a common characteristic in SCA patients, unlike in individuals with neurological diseases such as Parkinson's disease (PD) and Huntington's disease (HD), in which progressive protein-energy malnutrition is almost always present.^{21,22}

The exclusion of the two wheelchair patients may have contributed to the nutritional profile found, since both had a high SARA score (32 and a maximum value of 36.5).

Weight was only affected by both types of SCA in women. Women with SCA3 had a mean weight 16% lower than that of controls, while women with SCA10 had a mean weight 39% greater. In two studies with PD patients, reduced weight was also observed only in women. However, neither study managed to identify the reason for this.^{23,24} One of the authors suggested a possible association with hormonal and metabolic changes.²³ The lower weights observed in our SCA3 patients, even though within normal limits, may be due to the increase in energy expenditure caused by hyperkinesia (parkinsonism, dystonia, myoclonus, and chorea) reported in SCA3 but not in SCA10³; to dysphagia, which is more severe in SCA3 than in SCA10³; or to other unknown factors. Further research is required to investigate these changes.

In neurodegenerative diseases, involuntary weight loss is multifactorial. In PD, muscular rigidity and tremor are considered responsible for the increase in energy expenditure which, together with dysphagia, leads to weight loss^{25,26}. In HD, however, some studies have shown that weight loss is due to factors such as systemic metabolic changes caused by mutations in the huntingtin gene^{27,28} and the greater energy expenditure associated with involuntary movements (chorea) and rigidity, particularly in advanced stages of the disease^{21,23}. Although the characteristics of neurodegenerative diseases vary, according to all the studies referred to above weight loss is an important risk factor for increased comorbidity and mortality in these patients.

In the present study, height was one of the variables affected by the presence of SCA in both genders. Although lower stature can be attributed to a possible anatomical change in the spinal column of these patients, no studies corroborating this were found in the literature. It should be remembered that differences in stature were corrected by R in the FFM equation.

Only women with SCA10 were overweight. These women are therefore at greater risk for comorbidities associated with metabolic syndrome and reduced mobility. However, when patients with neurodegenerative diseases are assessed, BMI should not be analyzed separately because infiltration of fat into skeletal muscles in these patients due to myogenic atrophy leads to a higher fat-to-muscle ratio and can interfere with the results.²² In a study in which SCA3 patients were assessed, BMI was lower in both genders and was inversely correlated with trinucleotide repeat length.²⁹

Estimation of total body tissue composition by BIA confirmed the gender-related characteristics of body composition, i.e. BF and %BF are greater in women, and FFM is greater in men. The latter characteristic is well established and has been extensively discussed in the literature.^{30,31} Our confirmation of these characteristics was expected, as they are a consequence of the influence of sexual hormones on gender differences in body composition.^{31,32} Among the female subjects in this study, only those with SCA3 had reduced %BF and FFM. Female SCA10 patients, in contrast, although overweight, had %BF and FFM values similar to those of the controls. In a study of SCA1 patients, %BF was also similar in patients and controls.³³ As in the present study, this finding is probably due to the

BMI being at the upper limit of normality in the control group. With these results, the possibility that the SCA10 patients are developing sarcopenic obesity, as described by Petroni et al.,²⁵ in their study of PD patients, can be eliminated. Among the male subjects, there was also a reduction in both types of body compartments in SCA3 patients, while in SCA10 patients, there were only a reduction in FFM. In a study of HD patients by Süssmuth et al., although BMI was lower than in controls, FFM was similar. To explain this result, the authors put forward the hypothesis that these patients had lost BF.³⁴ Although there is statistically significant difference in disease duration between the male patients with SCA3 and SCA10, it is believed that there was no interference in the results, since the SARA scores are similar.

The present study is, to the authors' knowledge, the first to describe body composition characteristics in SCA3 and SCA10 patients of both genders, and the results should help to improve management of clinical and nutritional therapy in these patients. In a broad sense, this study has clarified the needs that must be taken in order to avoid body tissues losses that cause prejudices in the gait as well as possible comorbidities due to overweight.

The findings point to the need for distinct clinical and dietary therapies for each of the two forms of the disease, especially in females. Patients with SCA3 require hypercaloric and hyperprotein nutritional support, while in SCA10, the major focus should be on the control of sarcopenic obesity and its possible comorbidities. In addition, there must be biochemical research to detect both nutritional deficiencies and metabolic changes.

This study is an ongoing series of cooperative studies of both the Movement Disorders Division of the Neurology Service and the Nutrition and Dietary Department of the Hospital, on the nutritional status of SCA patients.

The small sample size, particularly that of the SCA10 sample, could be considered a limitation of this study. This limitation was compounded by the need to analyze male and female patients separately.

Finally, we concluded that SCA3 and SCA10 patients need nutritional follow-up to minimize body compartment changes.

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Table 1

Clinical characteristics of the SCA3 and SCA10 patients

Variable	Male		Female		* <i>p</i> value
	SCA3 (<i>n</i> = 13)	SCA10 (<i>n</i> = 6)	SCA3 (<i>n</i> = 19)	SCA10 (<i>n</i> = 8)	
Age (years)	46.2 ± 8.4	48.2 ± 8.9	44.5 ± 11.9	45.5 ± 7.5	0.907
Disease duration (years)	9.0 (6.0–11.0)	18.5 (14.0–32.0)	11.0 (8.0–14.0)	10.0 (4.5–15.5)	0.696
SARA score	10.0 (8.0–16.0)	9.2 (8.5–10.0)	13.0 (11.0–22.5)	8.7 (7.7–11.0)	0.051

Notes: SCA: spinocerebellar ataxia; n: number of individuals; SARA: Scale for the Assessment and Rating of Ataxia. Values are given as mean (± standard deviation) and median (first and third quartiles).
* Nonparametric Mann–Whitney test, *p*<0.05.

Table 2

Body composition of SCA3 patients, SCA10 patients and controls

Variable	Male				Female			
	SCA3 (13)	SCA10 (6)	Controls (35)	p-value*	SCA3 (19)	SCA10 (8)	Controls (41)	p-value*
W(kg)	71.4 (62.6–74.8)	73 (71.4–76.2)	72.8 (64.6–87.4)	0.479	52.0 (45.0–59.6)	69.9 (64.9–75.9)	62.0 (53.2–71.4)	0.001
H(cm)	170.1 ± 6.0	167.1 ± 5.0	173.7 ± 5.7	0.016	155.1 ± 5.9	157.4 ± 4.3	161.2 ± 6.9	0.004
BMI (kg/m ²)	23.4 ± 4.3	25.5 ± 2.6	24.7 ± 4.1	0.489	22.3 ± 3.8	28.5 ± 3.3	24.3 ± 4.2	0.002
BF(kg)	10.9 ± 5.1	13.7 ± 4.4	16.3 ± 8.0	0.081	12.7 ± 6.7	23.0 ± 5.1	19.7 ± 7.6	0.001
%BF	15.0 ± 6.1	18.7 ± 4.6	20.6 ± 7.1	0.047	22.4 ± 6.9	32.4 ± 4.9	30.1 ± 6.0	<0.001
FFM(kg)	50.6 (46.9–54.7)	51.2 (47.1–55.4)	58.6 (52.6–63.9)	0.002	38.2 (35.1–42.6)	45.0 (43.3–45.6)	42.8 (39.7–46.1)	0.005
PA	7.8 (7.0–8.2)	8.0 (7.6–8.2)	8.0 (7.7–8.3)	0.611	7.1 (6.6–7.8)	7.2 (7.0–7.7)	7.4 (7.1–7.7)	0.349

Notes: SCA: spinocerebellar ataxia; W: weight; kg: kilograms; H: height; cm: centimeters; BMI = body mass index; m: meters; BM: body mass; FFM: fat-free mass; PA: phase angle. Values are given as mean (± standard deviation) and median (first and third quartiles).

* One-way ANOVA, nonparametric Kruskal–Wallis test with 3 groups, $p < 0.05$.

Table 3

Anthropometric and bioelectrical impedance findings among SCA3 vs SCA10 patients and controls according gender

Variable	SCA3 vs. Control	SCA10 vs. Control	SCA3 vs. SCA10
Male			
<i>H</i> (cm)	0.057	0.012	0.295
% BF	0.014	0.515	0.268
FFM (kg)	0.001	0.008	0.990
Female			
<i>W</i> (kg)	0.006	0.039	<0.001
<i>H</i> (cm)	0.001	0.126	0.409
BMI (kg/m ²)	0.080	0.008	<0.001
BF	0.001	0.238	0.001
% BF	<0.001	0.334	<0.001
FFM (kg)	0.004	0.258	0.004

Note: SCA: spinocerebellar ataxia; *H*: height; cm: centimeters; BF: body fat; FFM: fat-free mass; kg: kilograms; *W*: weight; BMI: body mass index; m: meters. Student's *t* test or Mann–Whitney test, $p < 0.05$.

Table 4

Correlation between MM and ACS characteristics in groups without gender distinction

Groups	Variables	<i>n</i>	Correlation coefficient *	<i>P</i> value
SAC3	MM × Disease duration	32	−0.42	0.016
	SARA × MM	32	−0.38	0.033
	SARA × Disease duration	32	0.59	<0.001
SAC10	MM × Disease duration	14	0.25	0.389
	SARA × MM	14	0.11	0.697
	SARA × Disease duration	14	0.38	0.174

Note: *n*: number of patients; SAC: spinocerebellar ataxia; MM: muscle mass; SARA: Scale for the Assessment and Rating of Ataxia.

* Pearson or Spearman correlation coefficient $p < 0.05$.