

BODY COMPOSITION AND CHEST EXPANSION OF TYPE II AND III SPINAL MUSCULAR ATROPHY PATIENTS

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Abstract

Introduction: spinal muscular atrophy patients present muscle weakness, orthopedic problems, nutritional complications and respiratory impairment. Lean mass and fat mass modifications are also expected in this population. **Objective:** to verify the body composition and chest expansion of type II and III spinal muscular atrophy patients. **Methods:** fourteen individuals were evaluated: seven patients in Group I of 9 (7-12) years of age, weighing 29.7 (23.5-60.0) kg; and seven children without the disease in Group II of 9 (9-12) years, weighing 31.0 (27.8-54.1) kg. Patients' monofrequency bioelectrical impedance was used for analyze body composition. Chest, hip and abdominal girths were measured by a flexible steel tape. The SPSS program was used to statistical analysis ($p < 0.05$). **Results:** patients presented higher impedance: 1416.9 (850.5-1559.1) vs 788.0 (683.6-853.8), $P < 0.05$; and fat percentage: 31.2 (23.9-46.6) vs 19.1 (14.9-27.0)%, $P < 0.05$. The difference between forced inspiration and forced expiration thorax girth was smaller for patients when comparing to Group II: 3.0 (0.8-4.4) vs. 5.0 (3.9-6.5) cm, $P < 0.05$. **Conclusions:** patients with spinal muscular atrophy presented higher adiposity and lower chest expansion.

Key words: neuromuscular diseases; electric impedance; body fat distribution; anthropometry; lung.

INTRODUCTION

Spinal muscular atrophy (SMA) is a recessive, autosomal neuromuscular disease characterized by degeneration of anterior horn spinal cord motor cells and brain stem neurons¹⁻⁵. It is classified by disease severity and the age at onset of symptoms, namely type I for the most severe cases and type IV for those presenting few complications^{1,3,6-8}. Type II SMA infants get to three point of sitting independent and present thoracic deformity because of muscle weakness, which causes postural deviations. Type III SMA onset occurs in older children. Patients with type III get to three point of walking, whether or not they maintain this ability throughout adulthood^{1-3,6,7}. SMA patients present progressive symmetrical proximal weakness and hypotonia^{1-4,6,8}, but there is no sensory abnormality⁷. Besides muscle weakness, respiratory⁹, orthopedic⁵, and nutritional¹⁰⁻¹² problems are particularly noteworthy.

Among the methods used to evaluate body composition in children, bioelectrical impedance stands out as a noninvasive and painless method, which has already been adopted by other authors while evaluating SMA patients¹³, or children and adolescents without the disease¹⁴⁻¹⁶. Body girth measurements are frequently used in clinical practice^{17,18} due to their practical character and low price. Thoracic girth has already been associated with pulmonary function¹⁹, without any intention of replacing the standard spirometry measurements, used for a more frequent follow up of patients' pulmonary function.

SMA children suffer from stunted development due to nutritional, muscular, postural and respiratory alterations. Body composition assessment in these patients is proposed by Caromano et al.²⁰, since they state that there is a correlation between fat percentage and muscle strength in neuromuscular patients. There are few studies assessing neu-

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romuscular patients with more specific methods than bioelectrical impedance and anthropometric measurements. Therefore, the aim of this study is verify the body composition and chest expansion of type II and III spinal muscular atrophy patients.

METHODS

Study Protocol and Sample

Seven SMA patients (five with type II and two with type III) were recruited (Group I) from the neuropaediatric outpatient clinic of the Institute of Child Health and Pediatrics Martagão Gesteira (IPPMG), Federal University of Rio de Janeiro (UFRJ) and were evaluated at the Postgraduate Program of Rehabilitation Sciences laboratories, Augusto Motta University Center (UNISUAM). To compare data, seven individuals without the disease (Group II, control) had already participated in a previous research project at UNISUAM were selected from Rui Barbosa School (Bonsucesso – Rio de Janeiro, RJ), who had their age, sex, weight and height characteristics being paired with those of the patients (Table 1).

Assessments

Weight

Total body mass measurement was performed with an analog scale (R110, Welmy - Santa Bárbara d'Oeste, São Paulo, Brazil) to the nearest 0.1 kg. The subject was instructed to maintain both feet on the platform, without support and with the entire weight divided between both lower limbs²¹. For those that could not maintain the upright posture, this measurement was done with another person's help (supporting the patient) and subtracting the weight of the assistant from the total weight measured.

Height

Height was estimated for all participants (Groups I and II) by the equation proposed in the literature for Brazilian data²² because a great number of patients could not stand up and had lower limb deformities:

$$\text{Height (cm)} = 63.525 \\ 3.237 * (S) - 0.06904 * (A) + 1.293 * (HS)$$

Where S = sex (boys = 1; girls = 2); A = age (years) and HS = half span (cm, distance between the sternum and the distal phalange of the third finger, with the flexible steel tape parallel to the clavicle²³).

Bioelectrical Impedance

Body composition analysis was performed using a monofrequency bioelectrical impedance analyzer (BIA 310e, Biodynamics, Seattle, Washington, USA). Two electrodes were applied to the dorsal surface of the right hand and two electrodes were placed

on the dorsal surface of the right foot. The exam was performed in the supine position and the subjects received standardized recommendations: 1) no alcohol consumption or exercise within 24 hours prior to the test; 2) no caffeine or food consumption for four hours prior to the test; 3) the consumption of two-four glasses of water within the two hours prior to the test; 4) bathroom use within the 30 minutes prior to the assessment. Resistance and reactance values provided by the analyzer were used to estimate the fat-free mass (kg), in a child-specific equation^{14,15}: $FFM = 2.33 + 0.588 * (H^2/I) + 0.211 * W$, where FFM = fat-free mass (kg); H = height (cm); I = impedance ($Resistance^2 + Reactance^2$)^{1/2} (W); W = weight (kg)¹⁴. Fat percentage was calculated by the following equations: $FM = W - FFM$; and $\%F = (FM/W) * 100$, where FM = fat mass (kg), W = weight (kg), FFM = fat-free mass (kg), and % F = fat percentage (%).

Additionally, body mass index ($BMI = W/H^2$), fat-free mass index ($FFMI = FFM/H^2$) and fat mass index ($FMI = FM/H^2$) were calculated as previously described¹¹.

Girths

A flexible steel tape (Terrazul, Cambuci, São Paulo, Brazil) was used to measure the body girths. The abdomen measurement was made at umbilical level²⁴ and the hip girth, at the greatest girth, proximately to pubic symphysis²¹. For those that could not maintain the upright posture, measurements were taken with another person's help, supporting the patient's weight while the examiner performed the measurement. For thoracic girth, the flexible steel tape was placed at the level of the mesosternum, with arms abducted²¹. For those that could not maintain posture, the measure the upright was performed in a sitting position, preferentially without the use of chair back. Three measure were taken: 1) standard after a normal expiration (Thorax)²¹; 2) maximal inspiration after a forced inspiration (ThoraxInsp); and 3) maximal expiration after a forced expiration (ThoraxExp). The difference between maximal inspiration and maximal expiration girths was called 'chest expansion'.

Ethical Aspects

Informed consent was obtained from the childrens' parents and the protocol was approved by UNISUAM Ethics Committee before being applied (CAAE: 0028.0.307.000-11).

Statistical Analysis

Nonparametric descriptors and tests with resampling methods were selected because of the small sample size. Data were expressed as median (interquartile range). The Mann Whitney U test was used to compare groups, and the Monte Carlo simulation with 10,000 bootstrap samples was applied. All analyses were realized on the SPSS program, version 13.0, considering a 5% level of significance.

RESULTS

Table 1 summarizes demographic, body composition and girth variables separated by group. The age of Group I was 9 (7-12) years old, similar to Group II: 9 (9-12) years old (P = 0.509). The groups were also comparable for weight, height and BMI: 29.7 (23.5-60.0) vs. 31.0 (27.8-54.1) kg,

P = 0.618; 1.45 (1.40-1.58) vs. 1.55 (1.45 - 1.63) m, P = 0.262; and 15.3 (9.8 - 21.3) vs. 14.0 (12.9-18.8) kg/m², P = 0.905, respectively.

Group I presented higher values of resistance, impedance and fat percentage. Lower values of chest expansion, fat-free mass and FFMI were also observed, though the last two variables presented no statistical difference (Table 1, Figure 1).

Table 1: Demographic, body composition and girth variables

| Variables/Groups | Group I (n = 7) | Group II (n = 7) | P-value* |
|---------------------------|-----------------------|---------------------|----------|
| Half span (cm) | 65.1 (60.3-77.0) | 74.0 (66.0-80.0) | 0.171 |
| Resistance (W) | 1416 (850-1557) | 785 (681-849) | 0.005 |
| Reactance (W) | 51 (43-68) | 60 (54-70) | 0.267 |
| Impedance (W) | 1416.9 (850.5-1559.1) | 788.0 (683.6-853.8) | 0.008 |
| FM (kg) | 9.3 (5.7-27.7) | 5.3 (4.6-13.7) | 0.320 |
| FMI (kg/m ²) | 5.4 (2.7-11.1) | 2.6 (1.9-4.8) | 0.169 |
| FFM (kg) | 20.3 (16.0-26.7) | 26.4 (24.2-33.6) | 0.055 |
| FFMI (kg/m ²) | 9.5 (7.1-11.9) | 12.1 (10.9-12.9) | 0.055 |
| Thorax (cm) | 64.2 (56.5-79.1) | 63.2 (60.5-78.5) | 0.624 |
| ThoraxInsp(cm) | 69.0 (60.3-86.8) | 67.1 (66.0-79.0) | 0.757 |
| ThoraxExp (cm) | 63.5 (56.1-78.1) | 63.2 (59.5-77.5) | 0.624 |
| Chest Expansion (cm) | 3.0 (0.8-4.4) | 5.0 (3.9-6.5) | 0.049 |
| Abdomen (cm) | 60.5 (49.9-82.3) | 60.7 (57.0-75.0) | 0.945 |
| Hip (cm) | 72.0 (60.0-80.6) | 70.5 (66.0-81.0) | 0.811 |

Data are expressed as median (interquartile range). * Mann-Whitney U Test, with Monte Carlo simulation. FM = fat mass; FMI = fat mass index; FFM = fat-free mass; FFMI = fat-free mass index; Thorax = thorax girth after a normal expiration; ThoraxInsp = thorax girth after a forced inspiration; ThoraxExp = thorax girth after a forced expiration.

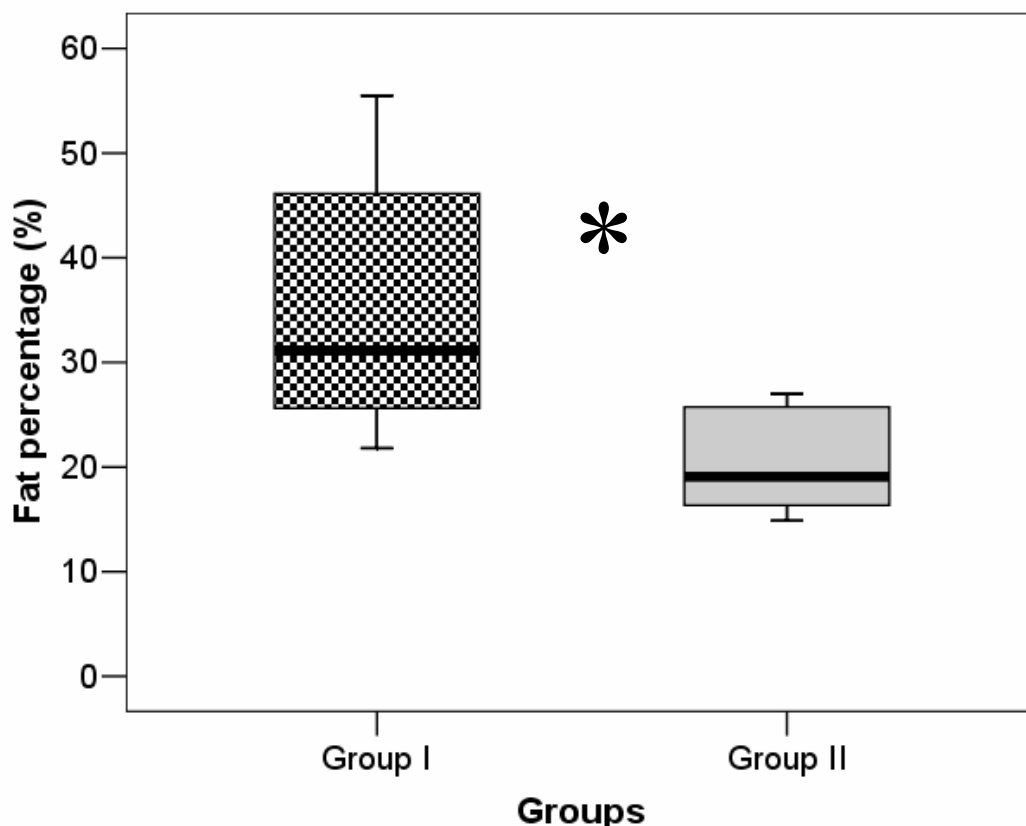


Figure 1: Groups I and II fat percentage box plot (minimum, 1st quartile, median, 3rd quartile and maximum). *P < 0.05 (Mann Whitney U test, with Monte Carlo simulation).

DISCUSSION

Group I composed of SMA patients exhibited higher body adiposity (with both high impedance and body fat percentage) than Group II. Leroy-Willig et al.²⁵ applied magnetic resonance and anthropometric measures to study the body composition of 11 children with neuromuscular diseases. Those authors observed an average of 35.9% of fat (skinfold thickness method), similar to the median values of the present study (31.2%). However, those authors included only three patients with SMA type II and did not investigate the thoracic expansion of their sample.

Higher values of body fat percentage and FMI as compared to regional normative data (paired by sex, age, and ethnic data) in the United States were also observed in 25 types I, II, and III SMA patients¹¹. Another American research group¹³ showed that 21 patients with types II and III SMA have higher levels of adiposity than subjects without the disease, considering either the thickness of subcutaneous fat tissue or the use of multifrequency bioelectric impedance. In agreement with this study, subcutaneous fat evaluated by a skinfold thickness caliper in 25 SMA patients (type II: 15; type III: 10 patients) was higher (type II: 12.5 mm; type III: 8.82 mm) than that of individuals without the disease (average of 5.37 mm) in the United States²⁶.

A study from Rio Grande do Norte, Brazil²⁷, assessed the fat percentage in 25 patients with neuromuscular diseases but presented results which cannot be compared with ours. In that study, 48% of the sample presented with optimal values, the remainder being categorized as 'low' and 'very low'. However, used the different method for assessment of fat percentage between these studies (skinfold thickness technique vs bioelectric impedance analysis) must be falceer into consideration, as well as the fact that only four SMA patients were included in the heterogeneous group of patients with neuromuscular diseases.

In a recent update, Markowitz et al.³ stated that SMA patients have excessive fat mass in relation to their muscle mass even for patients with normal BMI, as observed in our Group I. This fact may suggest to the nutritional health professional, who may assess the SMA patient only by BMI, to prescribe a dietary program to increase weight⁶ that he should this reinforcing the fat accumulation in this population. These findings justify the use of more precise measures of body composition in SMA patients. Overweight occurs mainly in type II, III, and IV⁶ SMA patients because type I patients present bulbar complications which cause an abrupt reduction of body weight (e.g., swallowing difficulties). The physiological process by which the fat accumulates in this population is not yet clear, a possible hypothesis being the replacement of myocytes by both conjunctive and adipose tissue in advanced stages of this chronic disease.

The fat-free mass in Group I was lower through without statistical significance than in the healthy group. This small amount of muscle mass corroborates the clinical profile of weakness observed in SMA patients^{1-4,6,8}. A recent Brazilian study²⁷ reported that 90% of the patients exhibited severe depletion of muscle reserve, in agreement with our results. The fact that these patients present low levels of physical activity due to muscle weakness and contractures reinforces the low energetic expenditure and the risk of obesity⁶. The increased fat mass represents an additional threat to the already compromised muscles that may lead to diminished motor function¹¹.

It is worth noticing that the height as estimated from the equation used in this study¹⁴ was used to calculate the fat-free mass suggested by Rabito et al.²². Those authors observed a coefficient of determination equal to 0.88 and considered this value between very good and excellent²⁸, suggesting that it was a valid strategy to apply this formula to all subjects (including those that could stand up for measurement of height). There is a consensus in the literature concerning the difficulties in obtaining accurate height measurements due to muscle contractures or inability to stand up, the use of measurements of other body segments and the half span (as in the present study) being suggested to estimate the patient's height⁶. The average arm span of three SMA patients was 140 cm in a previous study²⁵ and is similar to the double of the median half span observed in the present study (67.87 cm x 2 = 135.74 cm).

Data from a previous study²⁹ on pulmonary function (spirometry) in SMA patients showed a restrictive disorder characteristic of this population as resulting from respiratory muscle weakness, and reduced both chest wall and pulmonary compliance. Small values of thoracic expansion in the studied sample confirm this respiratory restriction and show that assessments by a simple instrument (such as a flexible steel tape) can be used for the follow-up of pulmonary function. According to Lima and Lima³⁰, girth measurements in children are important since it is a simple method, thus allowing the follow-up of bodily dimensions in clinical practice. A characteristic feature of type II SMA patients is the major weakness of the intercostal muscle, in addition to weakness of the other inspiratory and expiratory muscles⁶. The observed difference in thoracic expansion for Group I corroborates the above-cited relationship in a recent review about SMA³, when the respiratory muscle weakness, the restrictive pulmonary function, and the natural history of disease are considered.

In conclusion, type II and III SMA patients present higher adiposity and lower thoracic expansion as compared to a paired sample without the disease.

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