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Original article

Clinical assessment underestimates fat mass and overestimates resting energy expenditure in children with neuromuscular diseases

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SUMMARY

Background: Nutritional problems are frequent among patients with neuromuscular diseases, who consequently need an adequate evaluation. Objective: to describe nutritional assessment and to estimate and measure body composition and energy requirement in children with neuromuscular diseases.
Subjects and methods: We performed anthropometry, skinfold measurement and bioelectric impedance analysis (BIA) for estimate and measure, respectively, fat mass (FM). Resting energy expenditure (REE) was estimated by Schofield equations and measured by indirect calorimetry (IC). We compared actual energy intake with post-assessment recommendations.
Results: We studied 40 patients, 13.6 ± 3.3 years old (Range: 5.8–19.3), 80% boys, diagnosed with Duchenne Muscular Dystrophy (n = 21), other dystrophies (7), Muscular Spinal Atrophy (7), myopathies (3) and others (2). According to body mass index (BMI) 22.5% were well nourished (zBMI −1 to +1), 17.5% overweight (zBMI +1 to +2), 17.5% obese (zBMI ≥ +2), and 42.5% undernourished (zBMI < −1). Estimated FM was 20.2% (3.6–46.3), lower than BIA measurement: 34.2% (9.6–60.5) p < 0.001. Estimated REE was higher than measured REE: 1325 (813–2244) vs. 1202 (900–2100) kcal/day, p = 0.002. Actual energy intake: 1452 (1033–2476) was higher than recommended: 1300 (900–1900) kcal/day, p < 0.001.
Conclusion: Undernutrition and overweight are prevalent in this group of children with neuromuscular diseases. Clinical assessment underestimates FM and overestimates REE.

1. Introduction

Patients with neuromuscular diseases (NMD) are a heterogeneous group that presents improved survival rates mainly associated to multidisciplinary treatment and to the development of new technologies and methods of support [1]. Even though the incidence of NMD ranges from 1 in 3500 live male newborns for Duchenne Muscular Dystrophy (DMD) to 1 in 6000 for spinal muscular atrophy [2], their increased survival challenges the professionals involved in their medical care.

Nutritional management is an important aspect in overall treatment of NMD patients, given its impact in their prognosis and quality of life. Nevertheless, nutritional problems remain underdiagnosed; up to 56% of children with neurological disabilities develop undernutrition, while overweight is less frequently reported [3,4]. Both conditions, deficit and excess, increase morbidity and mortality, limit their ability to undertake daily activities and affect pulmonary function, a significant aspect that could be optimized by improving nutrition [1].

Two frequent areas of concern in nutritional care of NMD patients are: to perform an adequate nutritional assessment and to evaluate their energy needs. Due to their distinct body composition (BC) and reduced physical activity, these patients have low energy requirements, so measuring resting energy expenditure (REE) through indirect calorimetry (IC) can be very useful to guide their nutritional recommendation. However, IC is not always available, making it necessary to estimate REE through equations. On the other hand, given the low correlation between body mass index (BMI) and lean body mass (LBM) [13], evaluation of BC enriches anthropometric assessment. The basic clinical tools used in evaluating BC are

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skinfold measurement to estimate FM, and brachial perimeter to estimate LBM. Both have limitations, so it is necessary to perform specific tests, among which the bioelectric impedance analysis (BIA) is a simple test to measure BC, has better repeatability than skinfolds and good correlation to the reference method [5].

Our hypothesis was that bedside nutritional evaluation is possible in children with neurodissabilities, in the absence of sophisticated assessment tools. Our main objective was to describe the nutritional status of a group of children and adolescents with NMD, by estimating and measuring their body composition and energy requirements, in order to provide an appropriate dietary energy recommendation adapted to their particular situation.

2. Subjects and methods

2.1. Patients

We carried out a descriptive observational study; we invited 60 families and their children or adolescents with NMD from the National Program of Home Non-Invasive Ventilatory Assistance. Forty patients agreed to participate. A physician (SB) and a trained pediatric resident (REP) evaluated the patients in an outpatient facility from Josefina Martínez Hospital. We registered the patient's demographic data, diagnosis, physical activity with a functional motor scale [6], ventilatory support, previous nutritional assessments and a 24-h diet recall. Patients had a 12-h fast and were evaluated at the Nutrition Laboratory of the Pontificia Universidad Católica de Chile Hospital.

2.2. Anthropometry

Weight was measured with a Seca® standing scale or hammock scale according to the patient's functional capacity, and was expressed in kg with one decimal. If the subject could stand-up, we measured height (in m) with a stadiometer, or in supine position, on a firm surface when it was not possible. In this case, with a non-extendable measuring tape and when it was not possible to reach full extension, we measured by segments. This method has limitations: although is not validated, it is frequently used in clinical care considering the difficulties to perform an accurate assessment of stature in those patients. Equations for estimate length from particular segments are validated only for children with Cerebral Palsy, and its reference values for normal children is not easily acceded. The measurement was done by one of the authors.

Anthropometric indexes: height/age (H/A) and BMI (Weight in kg/Height [2] in m), according to CDC-NCHS reference [7] were expressed in percentiles and z scores (z score = [(real − ideal)/1 SD]). Patients were classified as: undernourished (zBMI < −1), well nourished (zBMI −1 to +1), overweight (zBMI +1 to +2) and obese (zBMI ≥ +2). Height/Age was classified as: normal (zH/A −2 to +2), Low (H/A < −2) and High (H/A > +2). Skinfold thickness measurement was performed by one experienced author (SB) with a Lange® caliper (Cambridge Scientific Instruments, Cambridge, MD) in four sites: Bicipital, tricipital, subscapular and suprailiac. Brachial perimeter (BP) was measured with a non-extendable measuring tape, all according to standard technique [8].

2.3. Body composition (BC)

Fat mass (FM) percentage was estimated with Slaughter equations [9]. LBM and body water were measured with a tetrapolar bioimpedance analyzer (Bodystat 1500), FM was calculated by the difference between total body weight and measured LBM.

2.4. Resting energy expenditure

REE was estimated using Schofield equations, considering age, sex and weight [10], expressed in kcal/day. Indirect calorimetry (IC) was performed with a Deltatrac II® metabolic monitor, measuring inhaled O_2 and exhaled CO_2 volumes, calculating the respiratory coefficient and REE. The equipment was calibrated before every measurement and was serviced annually. The steady state was established automatically and was also hand-calibrated selecting values with a variation coefficient lower than 6%. RQs were in a physiologic range.

2.5. Nutritional recommendations

Individualized recommendations were prepared according to the assessment and the physical activity level. Daily energy requirement was calculated with the equation used for patients with neurological illnesses, considering measured REE and corrective factors: muscle tone, physical activity (bedridden, wheel chair dependent, crawling or ambulatory) [6] and growth. We compared it to the actual intake (24-h diet recall) and to the recommended daily allowance (RDA) [10].

2.6. Ethics

The Ethics Committee of the Faculty of Medicine from The Pontificia Universidad Católica de Chile approved the study; parents and children older than 12 years of age signed an Informed Consent Form.

2.7. Statistical analysis

For statistical analysis we used the SPSS program, version 15.0 (SPSS. Inc. Chicago, IL). Results were expressed as median and range, or mean and standard deviation (SD), according to variable's distributions. We used non-parametric tests for group comparison: Wilcoxon test for dependent variables, Mann–Whitney test for the independent ones, the Kruskal–Wallis for more than two groups, and the Spearman correlation coefficient for simple linear correlation analysis. Significance was considered at $p < 0.05$.

3. Results

3.1. General characteristics

Forty patients were recruited, with a mean age of 13.6 ± 3.3 years (range: 5.8–19.3), 80% were male. Most of them had Duchenne Muscular Dystrophy (DMD, n = 21), followed by Spinal Muscular Atrophy (SMA, n = 7), other dystrophies (n = 7), myopathies (n = 3) and others (n = 2). 57.5% of the patients had non-invasive ventilatory assistance, during the night. With regard to functional motor capacity, 60% used wheel chair, 10% were able to drag or crawl and 30% needed assistance to walk. Only 42.5% (n = 17) reported having a nutritional assessment during the last six months, 27.5% (n = 11) at some point in the past, and 30% (n = 12) never had one.

3.2. Nutritional assessment and body composition

According to BMI, 22.5% of the patients were well-nourished, 42.5% undernourished, 17.5% overweight and 17.5% were obese. From all the sample, 23 (57.5%) had Low Height/Age (z H/A < −2). DMD patients had higher zBMI than no-DMD patients (ANOVA, p = 0.003).