Pediatric Neurology 52 (2015) 82–87

Contents lists available at ScienceDirect

Pediatric Neurology

journal homepage: www.elsevier.com/locate/pnu

Original Article

A Bedside Measure of Body Composition in Duchenne Muscular Dystrophy

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ABSTRACT

BACKGROUND: In clinical practice, monitoring body composition is a critical component of nutritional assessment and weight management in boys with Duchenne muscular dystrophy. We aimed to evaluate the accuracy of a simple bedside measurement tool for body composition, namely bioelectrical impedance analysis, in boys with Duchenne muscular dystrophy. **METHODS:** Measures of fat-free mass were determined using a bioelectrical impedance analysis machine and compared against estimations obtained from a reference body composition model. Additionally, the use of raw impedance values was analyzed using three existing predictive equations for the estimation of fat-free mass. Accuracy of bioelectrical impedance analysis was assessed by comparison against the reference model by calculation of biases and limits of agreement. **RESULTS:** Body composition was measured in 10 boys with Duchenne muscular dystrophy, mean age 9.01 ± 2.34 years. The bioelectrical impedance analysis machine values of fat-free mass were on average 2.3 ± 14.1 kg higher than reference values. Limits of agreement (based on 95% confidence interval of the mean) were −7.4 to 2.9 kg. There was a significant correlation between the mean fat-free mass and difference in fat-free mass between the bioelectrical impedance analysis machine and the reference model ($r = -0.86$; $P = 0.02$) suggesting that the bias was not consistent across the range of measurements. The most accurate predictive equation for the estimation of fat-free mass using raw impedance values was the equation by Pietrobelli et al. (mean difference, −0.7 kg; 95% limits of agreement, −3.5 to 2.0 kg). **CONCLUSIONS:** In a clinical setting, where a rapid assessment of body composition is advantageous, the use of raw impedance values, combined with the equation by Pietrobelli et al., is recommended for the accurate estimation of fat-free mass, in boys with Duchenne muscular dystrophy.

Keywords: duchenne muscular dystrophy, bioelectrical impedance, body composition, fat mass, fat-free mass

Pediatr Neurol 2015; 52: 82-87

Introduction

Duchenne muscular dystrophy (DMD) is the most common of the genetically inherited neuromuscular diseases in men, affecting one in every 3500 live male births.[1] The disease process follows a predictable course, altering body composition via progressive muscle wasting and degeneration resulting from the replacement of muscle with fat and fibrous tissue.[2] The body composition changes observed in DMD are unique; therefore, it is vital that accurate and acceptable techniques to assess body composition and body composition change in boys with DMD are available to clinicians which could be used to monitor disease progression.

Although laboratory-based body composition methods such as hydrostatic weighing, isotope dilution, and multicompartment models are often more accurate, each has inherent practical limitations, which render them unsuitable for routine use in clinical practice. Ideally, body composition measurement techniques in children with

Article History:
Received March 27, 2014; Accepted in final form August 6, 2014

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0887-8994/$ - see front matter © 2015 Elsevier Inc. All rights reserved.
http://dx.doi.org/10.1016/j.pediatrneurol.2014.08.008

chronic diseases need to be quick, noninvasive, and acceptable for repeated measures. The prediction of total body water (TBW) by bioelectrical impedance analysis (BIA) and in turn, body composition, is an inexpensive technique, which has applicability across a range of chronic diseases where standard body composition models are inaccurate.[3,4] Diseases such as juvenile rheumatoid arthritis and Crohn disease, in which chronic inflammation and subclinical malnutrition combined with use of corticosteroids is an example where BIA with disease-specific equations has proven to be a useful means of tracking nutritional status over time.[5,6]

BIA measures the impedance of the body to the flow of an alternating current.[7] The intracellular and extracellular fluids offer resistance to the flow, whereas cell membranes act as capacitors and thus offer reactance to the flow. As a result, impedance can be directly related to TBW. BIA is utilized in a range of clinical conditions to routinely measure body composition. Clinical trials have revealed the use of BIA as a noninvasive diagnostic tool to evaluate nutritional status, determine the prognosis of clinical patients, and evaluate the influence of therapeutic agents in disease management.[4,8-10]

Currently, there is limited information to inform and direct nutritional intervention in boys with DMD. The goal of weight management in children with this chronic disease is to preserve fat-free mass (FFM) while managing excess weight gain. However, standard anthropological measures such as body mass index (BMI) and skinfold measures provide only blunt measures and are invalid for use in this population.[11-14] Boys, who may appear "normal weight" according to their BMI, may have significantly increased fat mass. Similarly, changes in body composition as a result of disease progression or therapeutic treatment may not be observed using just BMI as an indicator or nutritional status. Assessment of body composition variables such as fat mass and FFM is consequently fundamental components of clinical management in boys with DMD. Furthermore, although the early introduction of corticosteroid treatment has led to significant improvements in physical ability and pulmonary function, side effects such as weight gain and changes in body composition require immediate and ongoing attention from clinicians.

Little is known about the use of BIA and its ability to accurately assess body composition of boys with DMD, particularly in those who now commence steroids very early in life. Furthermore, the anomalous body composition changes associated with DMD may alter body water distributions and consequently may negate the basic assumptions made in the BIA calculations.

As body composition in boys with DMD is of interest to clinicians and dietitians, as an indicator of disease progression, and/or management success, it is vital that techniques used to measure body composition are validated accordingly in boys with DMD. Consequently, the aim of this study was to evaluate the use of a clinical tool such as BIA for the estimation of FFM against estimations obtained from a reference three-component (3C) model.[15] Additionally, the use of raw bioelectrical impedance values was analyzed using three existing FFM predictive equations[16-18] for the estimation of FFM in steroid-treated ambulatory boys with DMD.

Materials and Methods

Patients

Ambulatory boys with DMD were recruited from two neuromuscular clinics in Australia ("MontroseAccess," a community center providing therapies for boys with DMD, and the Children's Neuroscience Centre at the Royal Children's Hospital, Melbourne). Diagnosis was defined as documentation of a deletion or duplication in the dystrophin gene, or absence of dystrophin on muscle biopsy, in conjunction with phenotypic evidence based on characteristic clinical symptoms or signs by 9 years of age (i.e., proximal muscle weakness, waddling gait, and Gowers maneuver), an elevated serum creatine kinase, and ongoing difficulty with ambulation. All boys were receiving corticosteroid treatment (prednisolone 0.12-0.65 mg/kg/day or deflazacort 0.83 mg/kg/day).

Ethics

The experimental protocol was approved by the Royal Children's Hospital Brisbane (2007/119), the Royal Children's Hospital Melbourne (29075B), and the University of Queensland Human Ethics Committee (2007000797). Written informed consent from parents and assent from the child were obtained before the commencement of the study.

Anthropometry

Height was measured to the last completed mm using a wall-mounted stadiometer (Holtain Instruments Limited, Crymych, United Kingdom), and weight was measured to the nearest 0.05 kg using calibrated electronic scales (Tanita BWB-600; Wedderburn Scales, Australia). BMI was calculated as weight divided by the square of height (m). Height, weight, and BMI were converted to *z* scores using Centers for Disease Control and Prevention reference values for children.[19] Pubertal status was recorded by a pediatric endocrinologist as Tanner stages.[20]

Body composition

Bioelectrical impedance was measured using a hand-to-foot multifrequency tetrapolar device (BodyStat 1500 MD; BodyStat, Isle of Man, United Kingdom), adhering to standard operating procedures with the subject's sex, age, height, and weight entered into the device, which enables FFM to be directly calculated from the internal algorithm (the default equation being Houtkooper et al.[21]). FFM was further calculated directly from the impedance index (ZI) as described by Kushner et al.[22] The ZI was then used to calculate FFM, using three different equations[16-18] that were derived from children with a similar age range as our study group (Table 1).

TABLE 1.
Predictive Equations for the Estimation of Fat-Free Mass

Source	Sex	Age Range (yr)	Equation
Bedgoni et al.[16]	M/F	7-13	FFM = 4.8 + 0.7ZI
De Lorenzo et al.[17]	M/F	7-13	FFM = 2.330 + 0.588ZI + 0.211WT
Pietrobelli et al.[18]	M	7-14	FFM = 0.6375ZI + 5.9913

Abbreviations:
F = Female
FFM = Fat-free mass (kg)
HT = Height (cm)
M = Male
WT = Weight (kg)
ZI = Impedance index