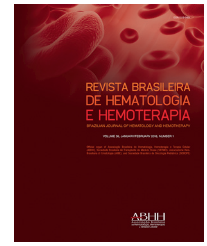
REV BRAS HEMATOL HEMOTER. 2017;xxx(xx):xxx–xxx

Associação Brasileira de Hematologia, Hemoterapia e Terapia Celular

Revista Brasileira de Hematologia e Hemoterapia
Brazilian Journal of Hematology and Hemotherapy

www.rbhh.org



Original article

Body composition of Fanconi anemia patients after hematopoietic stem cell transplantation

Priscilla Peixoto Policarpo da Silva, Daniella Schmit, Carmem Bonfim, Denise Johnsson Campos, Estela Iraci Rabito, Regina Maria Vilela *

Hospital de Clínicas da Universidade Federal do Paraná (HC/UFPR), Curitiba, PR, Brazil

ARTICLE INFO

Article history:
Received 31 March 2017
Accepted 9 June 2017
Available online xxx

Keywords:
Fanconi anemia
Body composition
Hematopoietic stem cell transplant

ABSTRACT

Introduction: Fanconi anemia is a rare genetic disease linked to bone marrow failure; a possible treatment is hematopoietic stem cell transplantation. Changes in the nutritional status of Fanconi anemia patients are not very well known. This study aimed to characterize body composition of adult, children and adolescent patients with Fanconi anemia who were submitted to hematopoietic stem cell transplantation or not.

Methods: This cross-sectional study enrolled 63 patients (29 adults and 34 children and adolescents). Body composition was assessed based on diverse methods, including triceps skin fold, arm circumference, arm muscle area and bioelectrical impedance analysis, as there is no established consensus for this population. Body mass index was also considered as reference according to age.

Results: Almost half (48.3%) of the transplanted adult patients were underweight considering body mass index whereas eutrophic status was observed in 66.7% of the children and adolescents submitted to hematopoietic stem cell transplantation and in 80% of those who were not. At least 50% of all groups displayed muscle mass depletion. Half of the transplanted children and adolescents presented short/very short height for age.

Conclusion: All patients presented low muscle stores, underweight was common in adults, and short stature was common in children and adolescents. More studies are needed to detect whether muscle mass loss measured at the early stages of treatment, results in higher risk of mortality, considering the importance of muscle mass as an essential body component to prevent mortality related to infectious and non-infectious diseases and the malnutrition inherent to Fanconi anemia.

* *Corresponding author at:* UFPR, Jardim Botânico Campus, Department of Nutrition, Av Prefeito Lothário Meissner n° 3400, Jardim Botânico, 80050-540 Curitiba, PR, Brazil. Tel.: +55 41 991115454.
E-mail address: regina.vilela@mail.mcgill.ca (R.M. Vilela).
http://dx.doi.org/10.1016/j.bjhh.2017.06.004
1516-8484/

Introduction

Fanconi anemia (FA) is a rare recessive genetic disease, usually inherited in an autosomal recessive manner, linked to bone marrow failure and an increased risk of developing a tumor.[1] FA patients are more prone to presenting morphological and endocrine abnormalities, such as bone deformities, cardiac and kidney malformation, hyperpigmentation, glucose intolerance, changes in glycemic control, dyslipidemia, hypothyroidism and growth hormone deficiency.[2]

The only hematology treatment that offers a potential cure for this disorder is hematopoietic stem cell transplantation (HSCT), which aims to restore the impaired bone marrow.[1–3]

Some conditions may affect the success rate of HSCT, such as the stage of the disease, the type of transplant, the origin of donor stem cells and histocompatibility, the conditioning regimen, age, previous treatment, and nutritional status of the patient.[4] Moreover, immunosuppression and the toxicity of the conditioning regimen have negative effects, in particular in respect to infections, bleeding, constipation, diarrhea, mucositis, nausea, and emesis.[3]

In addition to the characteristic outcomes of the disease, such as short stature, transplanted patients with FA can be underweight, present increased growth problems and dyslipidemia, whereas changes in glycemic and hormone control may result in an overweight condition or even obesity.[5]

Malnutrition has substantial prognostic and socioeconomic implications for patients and caregivers. Consequences of malnutrition include increased complications after surgeries and prolonged hospitalization resulting in higher exposure to infectious agents, a reduced response to treatment, a poorer quality of life and ultimately a worse prognosis. Individually, methods of nutritional assessment are limited and a gold standard has not been established to date. Consequently, at least two instruments have been used to establish the nutritional diagnosis adequately.[6] In the case of diseases that have complex metabolic demand and require a drug regimen that substantially affect cell structure and impose catabolism such as chemotherapy, the attention to nutritional status should be stressed, however, the criteria to choose the most adequate methods have not been clearly defined.[7]

As FA patients are more prone to malnutrition not only due to treatment but also because of the metabolic burden related to the disease, and as they might have higher risk of mortality, this study investigated whether changes in body composition are evident after transplantation and whether there are differences between patients submitted to transplant and those who are not. There are few studies that analyze the nutritional status of these patients, which makes it difficult to discriminate this group from patients with hematological diseases and as a consequence, during the clinical practice, nutrition support may not be specified to achieve their needs resulting in chronic malnutrition. Studies to better characterize the nutritional status of FA patients after HSCT are needed to optimize supportive care in this unique population.

The present study aimed to characterize the body composition of children, adolescents and adult patients with FA 1 year or longer after HSCT as compared to patients undergoing clinical treatment in order to support future studies related to nutritional assessment.

Methods

This cross-sectional study was conducted with male and female FA patients who were 2 years of age or older and submitted to HSCT or not at the Bone Marrow Transplant Service (BMTS) of the Hospital de Clínicas da Universidade Federal do Paraná. Exclusion criteria were patients that had been submitted to HSCT within 6 months of the start of this study, presence of physical changes that could impair anthropometric assessments, and cognitive difficulties to read, understand and fill out questionnaires. To characterize the population, a structured questionnaire was used consisting of identification data (date of birth and gender) and clinical data (submission to and type of HSCT, time after HSCT, and comorbidities).

This study was approved by the Human Research Ethics Committee of the Hospital de Clínicas da Universidade Federal do Paraná (#347232140.0.0000.0096), and a written consent was obtained from all participants or their legal guardians.

Anthropometric assessment

Weight, height, arm circumference (AC) and triceps skin fold (TSF) were measured, and body mass index (BMI) and arm muscle area (AMA) were then calculated. The 15th percentile was used as a cut-off point for inadequacy.[8] For adult patients (≥19 years old) BMI were classified according to the World Health Organization.[9] To classify children and adolescents, the WHO Antro® program was used for children younger than 5 years old, and the WHO Antro Plus® program was used for those aged between 5 and less than 19 years old. Z-score values were used to classify weight for age (W/age), height for age (H/age), and BMI for age (BMI/age) ratios.[10]

Body composition and phase angle assessment

Bioelectrical impedance analysis (BIA) was performed using a tetrapolar BIA device Quantum 101 (RJL System®, Inc. USA) with a current of 800 μA and frequency of 50 KHz. It was applied according to the instructions provided by the guide for bioelectrical impedance analysis.[11] Phase angle was calculated from the arc tangent resistance/reactance (Xc/R) value, which was expressed in degrees after being multiplied by $180/\pi$.[12] Phase angle standardization was based on the following equation: observed phase angle (°) – mean phase angle for gender and BMI (°)/phase angle standard deviation for gender and BMI.[13]

As there is no reference for phase angle stratified by BMI under 18.5 kg/m^2, 18 individuals were not considered in this analysis. For adults, phase angle values below 5° were considered risk predictors for malnutrition and morbidity.[14]

Lean body mass (LBM) values were obtained using the Kushner equation for children between 4 and 10 years old, the Houtkooper equation for children and adolescents aged from 11 to 18 years old, and the Lohman equation for adults aged between 19 and 29 years. Patients who did not meet the age