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Clinical, nutritional and family conditions of children with sickle cell disease followed up in a referral hematology center: a descriptive study

Condições clínicas, nutricionais e sociais de crianças com doença falciforme acompanhadas em um centro de referência: estudo descritivo

Abstract

Objective: To describe the clinical, social and nutritional characteristics of children with sickle cell disease aged 5 to 9 years. ***Method:*** Cross-sectional study carried out in 2015-2016 with a random sample of 190 children receiving care at a reference hematology hospital in Rio de Janeiro, Brazil. ***Results:*** The majority of the children had the genotype Hb SS (65.3%), were diagnosed with SCD during newborn screening (91%) and began clinical treatment before six months of age (91.5%). Overweight and low stature were found in 4.2% and 6.2% of the children, respectively. With respect to the household socioeconomic conditions, most of the parents had a partner (60.5%), lived with up to half the minimum wage (55.8%), and the women quitted their jobs to take care of their children (52.4%). ***Conclusion:*** Children with sickle cell disease were diagnosed early; nearly one sixth of them were overweight; and 20% of the parents had income of up to one fourth of the minimum wage. Strengthening the health care network in SUS for individuals with SCD is of vital importance to mitigate the problems identified in this study.

Keywords: Sickle cell anemia. Hemoglobin S. Child. Nutritional status. Socioeconomic factors.

Resumo

Objetivo: Descrever as características clínicas, sociais e nutricionais de crianças com doença falciforme de 5-9 anos. ***Método***: Trata-se de estudo seccional em amostra aleatória de 190 crianças avaliada entre 2015 e 2016, cadastradas em um hospital de referência em hematologia do Rio de Janeiro, Brasil. ***Resultados:*** A maioria das crianças estudadas possuía o genótipo Hb SS (65,3%), teve seu diagnóstico na triagem neonatal (91%) e iniciou o acompanhamento médico antes dos seis meses de idade (91,5%). Excesso de peso e baixa estatura foram encontrados em 14,2% e 6,2% das crianças, respectivamente. Em relação às condições sociais da família, a maioria dos pais possuíam companheiros (60,5%), viviam com até meio salário mínimo (55,8%) e as mulheres deixaram seus empregos para acompanhar o tratamento da criança (52,4%). ***Conclusão:*** As crianças com doença falciforme foram diagnosticadas precocemente, aproximadamente um sexto delas estavam em excesso de peso e 20% declararam renda de até um quarto de salário mínimo. O fortalecimento da rede de

atenção no SUS às pessoas com DF é uma importante medida para mitigar os problemas identificados neste estudo.

Palavras-chave: Anemia Falciforme. Hemoglobina S. Criança. Estado Nutricional. Fatores Socioecônomicos..

INTRODUCTION

Sickle cell disease (SCD) refers to a group of hematological disorders characterized by the presence of hemoglobin S (Hb S), being considered a public health problem in Brazil. According to newborn screening estimates in Rio de Janeiro, the incidence of SCD is 1: 1,200 live births.[1]

Individuals with SCD manifest chronic hemolytic anemia and episodes of pain due to the vaso-occlusion process caused by the sickle cells, which impair or even hinder blood circulation. These individuals are also more vulnerable to infections, splenic sequestration, acute chest syndrome (ACS), among other health complications.[2]

Growth deficit and low weight for age in children with SCD are a public health issue in low- and average-income countries. The results of a systematic review on the nutritional status of children with SCD showed lower anthropometric indicators in these children when compared with the population in reference. However, there has also been an increasing prevalence of overweight in children with sickle cell disease in this population.[3]

Children with SCD change the family routine, having to deal with diverse situations during the treatment such as a greater number of medical visits, complications of the disease and hospitalizations. Mothers play an important role in providing care: many of them do not work outside the house because they have to give the attention that their children need and be continuously on the alert.[4]

In addition to the imminence of clinical problems, which affect individuals with SCD and their families, socioeconomic conditions have been described as a concern in the course of the disease, such as low educational level, low income and a higher frequency of this disease in black people, who present lower socioeconomic indicators compared with the population in general.[5]

Given the above, the aim of this study was to describe clinical and nutritional characteristics of children with sickle cell disease and the social conditions of their families.

METHODS

It is a descriptive observational study with cross-sectional design with random sample of children with SCD aged between five and nine years, living in the metropolitan region of Rio de Janeiro and registered at a reference hospital that provides care and treatment to people with SCD – the *Instituto Estadual de Hematologia Arthur de Siqueira Cavalcanti* [State Hematology Institute] in Rio de Janeiro (Hemorio).

The sample calculated in this study represents the target population of children with SCD living in the metropolitan region of Rio de Janeiro, Brazil. The selection of the children receiving care at the hospital considered an estimated conservative prevalence of 50% of socially vulnerable children, at 95% confidence level. A total of 190 interviews were conducted, out of 202 individuals that comprised the sample, with loss of 6%. Data were collected between June, 2015 and April, 2016 by previously trained interviewers.

Information about the child, household' social conditions, the head of the household, and housing were collected through a face-to-face questionnaire used during the interview with the children's parents or legal guardians. Information on the children included: (i) Sex; birth weight

(<2,500g: low;> 2,500g to <4,000g: appropriate;> 4,000g: fetal macrosomia); genotype of sickle cell disease as informed by the parents (hemoglobin SS-Hb SS, hemoglobin SC-Hb SC, hemoglobin Hb Sβ-thalassemia, and hemoglobin SD); diagnosis of the disease (through newborn screening tests, symptoms of the disease, others) and beginning of follow-up (at 6 months, 2 years old, after 2 years old); (ii) Clinical intercurrences reported by the parents: hospitalizations in the last year as a result vaso-occlusion crises or infections; acute chest syndrome, priapism and stroke; transfusion therapies and use of hydroxyurea (yes or no for answer); (iii) Weight measured using a digital scale (Tanita®) with a capacity for 150 kg. The stature was measured with a vertical anthropometer with 100 cm of amplitude and graduation of 0.1 cm. The body mass index (BMI) was calculated by dividing the child' weight by the squared height. To assess the nutritional status, the height / age (H/A) and BMI / age indices, expressed as Z score, were calculated by the AnthroPlus® - 2007computer software and classified according to the World Health Organization.[6]

The variables relating to the household' social conditions were: (i) Information about the parents or legal guardians – educational level (college graduation, high school, elementary school, and illiteracy); marital status (with or without a partner); self-declared skin color (white, yellow and indigenous, brown and black); about the mother – if she quitted or lost the job due to absences to care for her child (yes or no), and if the family received the *Bolsa-família* government financial aid (yes or no); (ii) Data about the head of the household – defined as that who is responsible for paying the most part of the expenses (sex and occupation; employed, unemployed, retired/social benefits); (iii) data on housing (owned or rented home); number of people living in the house (up to three and more than four persons); number of individuals under 18 years old (up to two and more than three residents); basic sanitation (regular garbage collection; public sewage services and water supply) (yes or no), and range of *per capita* household income, which was obtained by dividing the sum of the overall earnings by the number of individuals living in the house – up to ¼, ¼ to <½ , from ½ to <1 and >1 of minimum wage .

Data analysis

Data were entered twice into an Excel® spreadsheet by different persons. Simple descriptive analyses of absolute and relative frequencies were carried out through the R Studio computer software, version 3.13 (RStudio).

The study was approved by the Research Ethics Committee of HEMORIO, process number 366/14. All parents/guardians of the children read and signed the Free and Informed Consent Form.

RESULTS

Based on the answers to the questionnaire administered to the parents or legal guardians of 190 children, the following data were found: male predominance (54.2%), adequate birth weight (79,6%), Hb SS genotype (65.3%), SCD detected in neonatal screening (91.0%), and beginning of follow-up before the age of six months (92.0%). With regard to clinical

intercurrences, 24.8% of the children needed hospital care at least three times in the year before the interview due to vaso-occlusion crises or infections. There were reports of transfusion therapies in 56.1% of the children and use of hydroxyurea in 30.2% of them (Table 1).

Table 1. Diagnosis and clinical characteristics of children with sickle cell disease receiving care at a state reference hematology hospital. Rio de Janeiro, RJ, Brazil, 2016.

Variables	N	(%)
Sex	190	
Male		54.2
Female		45.8
Birth weight	162	
Low		11.7
Appropriate		79.6
Macrosomia		8. 7
Genotype	190	
Hb SS		65.3
Hb SC		27.4
Hb Sβ / Hb SD		7.3
Disease diagnosis	189	
Neonatal screening		91.0
Symptoms of the disease		7.4
Others		1.6
Beginning of follow-up (age)	188	
Before 6 months old		92.0
Up to 2 years old		4.8
More than 2 years old		3.2
Clinical intercurrences	190	
Yes		29.5
No		70.5
Hospitalizations due to vaso-occlusion crises	187	
Yes		15.3
No		84.7
Acute chest syndrome	190	
Yes		14.7
No		85.3
Hospitalizations due to infections	188	
Yes		9.5
No		90.5

Table 1. Diagnosis and clinical characteristics of children with sickle cell disease receiving care at a state reference hematology hospital. Rio de Janeiro, RJ, Brazil, 2016. (Cont)

Variables	N	(%)
Priapism (boys)	100	
Yes		4.8
No		95.2
Stroke	190	
Yes		2.6
No		97.4
Transfusion therapies	189	
Yes		56.1
No		43.9
Use of hydroxyurea	189	
Yes		30.2
No		69.8

The BMI/age ratio of the children subjected to nutritional assessment was normal for most of them (83.5%). The percentage of overweight and obese children was 14.2%, and the percentage of low stature was 6.2% (Table 2).

Table 2. Nutritional status of children with sickle cell disease receiving care at a state reference hematology hospital, Rio de Janeiro, RJ, Brazil, 2016.

Variables	n (176)	(%)
Body mass index for age (kg/m^2)		
Underweight		2.3
Normal weight		83.5
Overweight		14.2
Height for age (z score)		
Adequate		93.8
Low stature		6.2

The majority of the parents or guardians graduated from high school (49.2%) and lived with a partner (60.5%), declared themselves as being black and brown (43.2% and 40.5%, respectively), and quitted the job to care for their child (52.4%). About 60% of the households were occupied by up to three residents, and 64.2% of the families had up to two residents under 18 years old. With respect to the *per capita* household income, 55% of the families lived with up to half a minimum wage, and 38.8% of them were beneficiaries of the Brazilian income transfer program (*Bolsa Família*) (Table 3).

Tabela 3. Socioeconomic conditions of the families with children with sickle cell disease receiving care at a state reference hematology hospital. Rio de Rio de Janeiro, RJ, Brasil, 2016.

Variables	n	(%)
Educational level of the child's parents/guardians	189	
College graduation		6.3
High school		49.2
Elementary school		42.9
Illiteracy		1.6
Marital status of the mother/guardian	190	
With a partner		60.5
Without a partner		39.5
Color/race	190	
White. Yellow and Indigenous		16.3
Brown		40.5
Black		43.2
Mother had quitted the job or was fired due to absences to care for the child *	189	
Yes		52.4
No		24.9
Does not work outside the house		22.7-
Beneficiary of *Bolsa-família* financial aid	188	
Yes		38.8
Sex of the head of the household		61.2
Sex of the head of the household	190	
Male		58.9
Female		41.1
Occupation of the head of the household	186	
Employed		75.8
Unemployed		13.4
Aposentado/ benefícios sociais		10.8
Situation of the dwelling	190	
Owned		69.5
Rented		30.5
Number of people living in the residence	190	
Up to3		63.2
More than 4		36.8
Number of residentes under 18 years old	190	
Up to 2		64.2
More than 3		35.8
Renda domiciliar *per capita*	190	
Up to ¼ of minimum wage		20.0
¼ to < ½ of minimum wage		35.8
½ to <1 minimum wage		26.3
> 1 minimum		17.9

DISCUSSION

One of the most important characteristics observed in the children with SCD was having the genotype Hb SS, described as being of major clinical severity, predisposed to the occurrence of clinical complications, and having born with appropriate weight. It is worth noting that most of the children were diagnosed with SCD during the newborn screening, showing an early start of care of the children's health, before six months of age.

In 2001, after implementation of Decree n° 8.229, a screening survey for hemoglobinopathies in 99,260 newborns in Rio de Janeiro reported that 94,513 infants (95.2%) had normal hemoglobin levels.[7] Early diagnosis is vital in case of SCD because, in addition to early beginning of care and preventive actions, it results in a better quality of life and decreased child mortality.

Although the children in this study were born after implementation of the *Programa Nacional de Triagem Neonatal* [National Program of Neonatal Screening], 10% of the SCD diagnoses were made on the basis of the symptoms of the disease or else. Diagnoses made in Brazil through the *Sistema Único de Saúde (SUS)* [Unified Primary Healthcare System] in 2013, covered 83.1% of the children.[8] However, it is still a challenge to absorb early in the healthcare system children diagnosed with SCD in the neonatal screening test.

Ensuring early diagnosis of SCD is one of the guidelines of the *Política Nacional de Atenção às Pessoas com Doença Falciforme e de outras Hemoglobinopatias* [National Policy for the Care of People with Sickle Cell Disease and other Hemoglobin Pathologies] (Decree GM/MS n° 1.391, of August 16, 2005),[1] which has the objective of reducing morbidity and mortality of children with SCD.

The reported clinical intercurrences in the children studied were acute chest syndrome, hospitalizations due to vaso-occlusion and/or infections, and the need for transfusion therapies. The occurrence of ACA and frequent hospitalizations (more than three times) is associated with a more severe SCD, being one of the main causes of death in children with this disease.[9] Our results reinforce the need to provide preventive care to minimize clinical manifestations of SCD in the routine visits to the healthcare centers.

In a Brazilian survey carried out in 2008/2009, representative of the general population of children aged five to nine years, prevalence of overweight was three and half times higher than that found in the present study.[10] The increased prevalence of overweight and obesity has been a public health issue worldwide. [11]

In 2013, Chawla and collaborators found 22.4% of overweight and obese children and adolescents with SCD and low prevalence of underweight in the group studied.[12] A higher prevalence of overweight and obesity in children with SCD were also found in the present study.

In our study, the frequency of overweight in children with SCD was 14.2%, higher than the ones found in other studies with individuals with sickle cell disease.[13,14] In São Paulo, a study with children with SCD in routine health follow-up at the Federal University of São Paulo found 9.2% of prevalence of overweight.[13] The shift in the nutritional profile of children with SCD in Brazil seems to be consistent with the nutritional transition process in the country.[10] Yet, overweight has been a new concern with children with sickle cell disease because an increased body mass requires an increase of blood flow and an overload in the organs already impaired by the disease.

The percentage of low stature in children with SCD in the present study was similar to the prevalence found in the last national survey conducted 2008/2009, that is, 6.8%.[10] The literature review carried out by Jesus et al. (2018)[3] showed that in older studies higher rates of underweight were found in children with SCD, and especially in developing countries, including Brazil, prevalence rates were higher.[14]

An increase in the basal metabolic rate and the presence of anemia were described as major factors for the deficits of height and weight in individuals with sickle cell disease, but it is difficult to determine the magnitude of contribution of intrinsic factors. An early beginning of follow-up of children with SCD in reference healthcare units, with provision of therapeutic resources such as hydroxyurea[15] and blood transfusions, can be an important step toward improving the nutritional status. Children with adequate nutritional status have a better prognosis of SCD and less chance of hospitalizations and recurrent infections.[16]

In the sampled population of this study, the use of hydroxyurea (HU) to diminish complications from SCD was reported for 30% of the children. In Brazil, the *Protocolo Clínico e as Diretrizes Terapêuticas para Doença Falciforme*[17] [Clinical Protocol and Therapy Guidelines for Sickle Cell Disease] recommends the minimum age of three years for use of this drug. The debate about the use of HU is focused on the toxicity of this medication and the parents' decision to allow or not its use, because it requires more medical monitoring and laboratory tests. [15]

Socioeconomic factors interfere in the health care and in the prognosis of diseases. Among the variables used to describe the household characteristics, it was found that most families lived with up to half the minimum wage *per capita,* and the majority of the parents or legal guardians completed elementary school. In a study that investigated the deaths of 193 children with SCD, a low household income was present in 90% of the deaths.[18]

A little over half of the children's mothers reported that they had to quit their jobs. This condition is aggravated by the fact that nearly 40% of the women interviewed did not have a partner, which to a greater or lesser extent may affect the dynamics of these families. Caring for children with SCD affects the life of the families. Their socioeconomic situation can be worsened when the parents or guardians need to quit the job due to frequent intercurrences and hospitalizations.

One of the strengths of the present study is the fact that it has been carried out with a random sample, enabling inference for the studied population living in the metropolitan region of Rio de Janeiro, in addition to the low percentage of losses. A limitation was that it was conducted only with children monitored by the public primary healthcare system (SUS), because children being treated by the private health system were not considered in the design of this study.

CONCLUSION

Despite an early diagnosis and monitoring of children with SCD, it was found that they remain clinically vulnerable as they advance in age due to severe clinical intercurrences that occur. The percentage of overweight children is a matter of concern, considering that an inappropriate nutritional status, higher or lower than standard, has a severe impact on the course of the disease. Actions of care of children that live with SCD must be considered under a perspective of integrated care, encompassing both specialized assistance and health monitoring, aiming to the quality of life of these children and the achievement of their full potential of growth and development.

It is worth noting that the findings of this study about the follow-up of the children by their mothers indicated that they renounced the labor market. Thus, these aspects cannot be ignored in the health care of this group.

Strengthening the network of health care provided by SUS to individuals with SCD is vitally important to mitigate the problems identified in this study. Primary care plays a key role in promoting a whole health

care, which would entail a care service not only focused on the clinical manifestations of the disease but on the individuals themselves, on their context of life.

The social inequities experienced by this group amplify the suffering caused by the disease. Thus, the role of social and institutional support networks is of vital importance, as is the case of sickle cell disease associations, as well as intersectoral articulations towards the promotion of education, social assistance, employment, income and quality of life of these families.

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Contributors

Santos IN, Aguiar OB, Damião JJ and Rodrigues CSC participated in the design, data analysis and interpretation, revision and approval of the final version of the manuscript. Freitas ACT and Voll VM participated in data collection and interpretation.

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