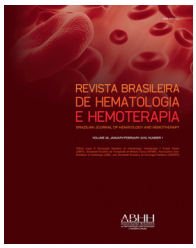
Revista Brasileira de Hematologia e Hemoterapia
Brazilian Journal of Hematology and Hemotherapy
www.rbhh.org



Original article

Oral health-related quality of life of children and teens with sickle cell disease

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ARTICLE INFO

Article history:
Received 24 June 2015
Accepted 19 January 2016
Available online 13 February 2016

Keywords:
Sickle cell
Quality of life
Child
Oral health
Malocclusion

ABSTRACT

Background: Children with sickle cell disease may have their quality of life affected by oral alterations. However, there is still little data on oral health-related quality of life in these children. The aim of this study was to investigate the influence of sickle cell disease, socioeconomic characteristics, and oral conditions on oral health-related quality of life of children and teens.

Method: One hundred and six children and teens with sickle cell disease were compared to a similar sample of 385 healthy peers. Data were collected through oral examinations, interviews to assess quality of life (Child Perceptions Questionnaire for children aged 8–10 and 11–14) and questionnaires containing questions on socioeconomic status.

Results: There were no statistically significant differences in the total scores of the Child Perceptions Questionnaires or domain scores comparing sickle cell disease patients to control subjects. When sub-scales were compared, oral symptoms and functional limitations had a greater negative impact on the quality of life of adolescents with sickle cell disease (p-value <0.001 and p-value <0.01, respectively) when compared to healthy controls. The only statistically significant determinants of negative impact on oral health-related quality of life in the overall sample was home overcrowding (more than two people/room) in the younger children's group, and dental malocclusion among teens.

Conclusion: There was no significant difference in the negative impact on the oral health-related quality of life between the group with sickle cell disease and the control group. Of the oral alterations, there was a significant difference in the oral health-related quality of life between adolescents with sickle cell disease and controls only in relation to malocclusion. Among the socioeconomic characteristics, only overcrowding was significantly associated with a negative impact on oral health-related quality of life.

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http://dx.doi.org/10.1016/j.bjhh.2016.01.004

Introduction

Sickle cell disease (SCD) is an inherited autosomal recessive blood disease. The inheritance of one sickle cell gene from each parent (SS) is the most common and the severest form of the disease, affecting around 280,000 newborns per year. This disease, in addition to thalassemia, is responsible for 3.4% of all deaths of children under five years of age.[1] Each year, 3500 children in Brazil are born with SCD.[2]

Children with SCD are at risk for serious morbidities related to vascular occlusion, hemolysis, and infection, which can impair their quality of life (QoL) and lead to early death. The pathological effects of SCD, seen in mineralized connective tissues, also occur in dental tissues and the oral cavity, usually in late childhood and during adolescence.[3] The most commonly described findings in the oral cavity, which are not pathognomonic but may be characteristic of the disease, are pallor of the oral mucosa due to a low hematocrit and depapillated tongue.[4] There are reports of delayed tooth eruption, hypoplasia and hypomineralization, hypercementosis, pulp stones and asymptomatic pulp necrosis due to thrombosis in the blood vessels.[5–7]

Individuals with SCD experience a lower QoL compared to healthy peers.[8,9] Due to the clinical course of the disease, SCD is thought to affect the QoL in multiple dimensions. The most serious organic changes result in emotional and physical stress for children and their families.[10] The frequency of episodes of fever, hospitalizations, and pain can trigger anger and sadness.[11] Moreover, lower health-related QoL in children with SCD is associated with a socioeconomic disadvantage,[12] a low level of education and not living with both biological parents.[13] Religion and spirituality have been identified by individuals with SCD as an important factor in coping with stress and in determining the QoL.[14]

QoL may be affected by oral conditions. Oral conditions such as dental caries and malocclusions affect self-esteem, the ability to chew and speak, and may be associated with absenteeism from school and psychological problems.[15,16] Although there have been studies on the QoL of patients with hematological diseases with regard to their behavioral and psychological impacts, emphasis on oral health has remained relatively underexplored. Only recently, the oral health-related QoL (OHRQoL) of 54 teenagers with SCD was evaluated by an adolescent medicine clinic in Columbus, Ohio, comparing them with adolescents with other chronic diseases. There was no statistically significant difference in the OHRQoL between the two groups.[17]

The objective of this study was to investigate the influence of SCD and factors related to the disease, oral conditions, resources and individual characteristics on the OHRQoL of children with this disease.

Methods

Ethical approval

This study received approval from the Ethics Committees of the Fundação de Hematologia e Hemoterapia do Estado de Minas Gerais (Hemominas) and the Universidade Federal de Minas Gerais (UFMG), Brazil. Written informed consent was obtained from the participants or parents/guardians of the participants of this study. This research was conducted in accordance with the Helsinki Declaration as revised in 2008.

Participants and recruitment

The study was conducted in the city of Belo Horizonte, the capital of the state of Minas Gerais, Southeast Brazil. The study sample was made up of children with SCD, residing in the metropolitan region of Belo Horizonte, aged from 8 to 14 years old, sampled from the patient registry of the referral center, Hemominas. A control group of healthy children and teens was recruited from the same schools attended by the children with SCD. A total of 450 children and adolescents with SCD aged 8–14 years were registered as receiving services from Hemominas in 2012. Among these, 196 children and teenagers resided in the metropolitan region of Belo Horizonte.[18]

The sample size was calculated from the expected standard deviations (SDs) of the QoL scales to evaluate the OHRQoL: the Child Perceptions Questionnaire for children aged 8–10 ($CPQ_{8–10}$) (SD = 10.7) and 11–14 years ($CPQ_{11–14}$) (SD = 10.1) investigated in a pilot study. The pilot study was conducted using a random sample of 34 children and 35 teens with SCD registered in Hemominas. The required sample size ($n = 51$ children aged 8–10 years, and $n = 45$ adolescents aged 11–14 years) was calculated based on the ability to detect a five-point difference in QoL scores on comparing SCD patients to apparently healthy controls, assuming an $\alpha = 0.05$ and $\beta = 0.10$. The program SPSS version 20.0 was used for all statistical analyses. One hundred and eighty healthy adolescents and 205 healthy younger children who were enrolled at the same schools and in the same classes as those with SCD were selected, and matched by age and gender. The option to match one case to at least every three controls was because healthy controls could be less motivated to participate than individuals in a health-care setting.[19]

Eligibility criteria for inclusion in the SCD group were as follows: diagnosis of SCD hemoglobin (Hb) SS in their medical records, not suffering from a painful crisis at the time of the survey, no medical conditions other than SCD, no emergency dental appointment within the previous three months and no intellectual disability. Eligibility of the members of the control group included: no organic, physiological, or psychiatric disorders and no intellectual disability, apparently healthy with no dental appointment within the previous three months.

Calibration exercise

Prior to the fieldwork, the examiner was calibrated and trained in respect to the diagnosis of oral diseases (dental caries and malocclusion). This consisted of two stages. The theoretical stage involved a discussion of the criteria for diagnosis of oral alterations. The second stage involved an oral clinical examination of children and adolescents not included in the study. Participants were examined in two sessions with a nine-day interval between. Data analysis involved the calculation of Kappa coefficients for inter-examiner agreement (malocclusion: $k = 0.91$; dental caries: $k = 0.92$; gingival bleeding: $k = 0.89$) and intra-examiner agreement (malocclusion: