

Health Related Quality of Life of Patients with Sickle Cell Disease

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Received date: Mar 11, 2016, Accepted date: Apr 01, 2016, Publication date: Apr 04, 2016

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Abstract

Objective: The aim of the present study was to evaluate the health-related quality of life in pediatric patients with Sickle Cell Disease.

Methods: An analytical cross-sectional study was performed. Pediatric patients suffering from Sickle Cell Disease below the age of 21 were compared with asymptomatic pediatric individuals. Clinical and demographic data were collected along with the Pediatric Quality of Life Inventory (PedsQL 4.0).

Results: The sample was composed of 68 children and adolescents in the "Sickle cell group" and 44 in the "Comparison group". Children and adolescent with SCD had lower scores of health related quality of life compared with healthy subjects in the domains physical (68.3 versus 88.8), social (71.1 versus 90.7), and school functioning (60.2 versus 78.7). The domain emotional functioning was not significantly different (62.1 versus 66.5).

Conclusion: The results of the present study demonstrated that children and adolescents with SCD had lower scores of quality of life in the domains activities, social and school functioning when compared to healthy pediatric individuals.

Keywords: Sickle cell disease; Quality of life; Evaluation

Introduction

Sickle cell disease (SCD) is the most common hereditary blood disorder in the world, and also the most prevalent in Brazil [1]. It is a severe chronic diseases associated with continuous treatment over an extended period of time, and significant human suffering [2]. It is well known that all chronic and acute complications of SCD could be expected to have a negative impact on quality of life of the patients, especially for children and adolescent [2,3]. Therefore, the evaluation of patients' health based almost exclusively on the clinical severity of the disease has certainly created distortions between the medical approach and expectation of individuals, especially in chronic diseases such as SCD [2-4]. Health-related quality of life (HRQoL) is a concept that may provide the subject's own view of his/her wellbeing and not the view of a health professional about the disease [4]. In spite of the importance of HRQoL in medical evaluation, few studies in this field have been focused on SCD, and rarer still are those that were dedicated to the pediatric age-range [2-5]. For this reason, the aim of the present study was to evaluate the health-related quality of life in pediatric patients with Sickle Cell Disease.

Patients and Methods

An analytical cross-sectional study was performed to evaluate quality of life of individuals suffering from SCD below the age of 21, compared with asymptomatic individuals originating from a public school at the same age. Patients were recruited from among those regularly seen at the Center for Hemotherapy and Hematology of the

State of Bahia, Brazil (HEMOBA) between February and November of 2011. This study was approved by the Ethics Committee of the Bahian School of Medicine and Public Health (protocol number 111/2010) and all children's guardians signed an Informed Consent Form.

The minimum sample size was calculated to detect a mean difference of five points, assuming a standard deviation of 17 [6] points and an alpha error of 0.05. The sample size was estimated at 44 subjects in each group. Patients (males and females) with a confirmed hematologic diagnosis of SCD between the ages of 8 and 21 years were eligible for enrolment in the study. The following patients were excluded: those with cognitive impairment; those with associated diseases not related to SCD such as rheumatic disease, etc. For the "Comparison group", children considered healthy and whose socio-demographic characteristics were similar to those of the "Sickle cell group" were selected.

Clinical and demographic data were collected from all the subjects. The Brazilian-Portuguese version of the Pediatric Quality of Life Inventory (PedsQL 4.0) was also applied^{13,14}. The PedsQL questionnaire 4.0 [4,7] was developed specifically for children in defined age ranges, and it was used for children in age intervals of 8 to 12 and 13 to 18 years. This study focuses on the health related quality of life of individuals as measured by the PedsQL self-report for children. Data was tabulated into distribution tables by frequency in the case of categorical variables or into mean and standard deviation in the case of continuous variables. For comparison between the groups with and without SCD, the chi-square test was used for discrete variables and the Student's t-test for continuous variables. All tests were 2-sided and P values less than 0.05 were considered statistically significant.

Results

The final sample was composed of 68 children and adolescents in the "Sickle cell group" and 44 in the "Comparison group". Social and

demographic characteristics are shown in Table 1. Comparison of the quality of life scores using PedsQL.4.0 demonstrated significant differences in the domains Physical, Social, and School (Table 2).

Characteristics	Sickle cell group	Comparison group	P
Age group	12,2 (3,3)	11,6 (3,2)	0.83
Weight	37.1 (± 12.5)	40.1 (± 13.7)	0.12
Height	1,4 (± 0,2)	1.5 (± 0.2)	0.01
Gender			0.09
Male	32 (47.1%)	28 (63.6%)	
Female	36 (52.9%)	16 (46.4%)	

Table 1: Main social and demographic characteristics of children suffering from sickle cell disease compared with health ones.

Quality of life domains	Sickle cell group	Comparison group	p
Physical Functioning	68,3 (18,3)	88.8 (± 7.3)	< 0.01
Emotional Functioning	62,1 (22,1)	66.5 (± 14.9)	0.25
Social Functioning	71,1 (23,5)	90.7 (± 12.6)	< 0.01
School Functioning	60,2 (20,7)	78.7 (± 16.1)	< 0.01

Table 2: Quality of life of children suffering from sickle cell disease compared with health ones.

Discussion

Sickle cell disease is the most common hereditary blood disorder in the world, and also the most prevalent in Brazil [1]. It is estimated that 3% of the Brazilian population are carriers of the sickle cell trait, and this number represents at least two million persons [1]. Therefore, the evaluation of the disease based on the patient's perspective is fundamental to the understanding of this severe chronic condition. In this group of patients, our findings showed that children and adolescent with SCD had lower scores of health related quality of life compared with healthy subjects. The most affected domains were physical, social, and school functioning.

SCD is a severe chronic diseases and the need for continuous treatment over an extended period of time is a factor which directly influence quality of life [1,5,6]. Acute complications of SCD includes painful episodes, acute chest syndrome, splenic sequestration, infection, stroke, aplastic crisis, and priapism. Common chronic complications include also pigment gallstones, delayed growth and development, bone avascular necrosis, pulmonary hypertension, and renal disease. It can thus be inferred that all those complications would be determining factors in quality of life, as cited in the literature [3,5,6].

Prior studies confirmed that children with SCD had significantly impaired health related quality of life when compared to healthy children [2,5,8]. The quality of life of our patients were not only lower than healthy children but also very similar to those found in patients suffering from other chronic conditions such as cancer receiving treatment and rheumatoid arthritis [2,5,8]. In all those studies, patients and caregivers have reported consistently more limited physical, psychological, and social well-being compared to healthy children.

Dale reported that 63% of the children with SCD are at risk for impaired physical health, 28% for social functioning and 55% for school functioning [2]. SCD is a chronic disease that causes deterioration in the health of the patients that requires frequent interventions and medical treatments [1,2,5,7,8]. These treatments may require hospitalization or treatment at home, in an ambulatory setting, or in the emergency department, thus affecting attendance at school and normal play activities [2]. Surprisingly, children with SCD did not differ significantly from the Comparison group in the domain emotional functioning. This is a light contrast to the results of previous findings [5,8]. However, we believe that children with SCD may have adjusted to their emotional deficits and therefore do not subjectively experience problems in these domains.

One of the limitations of this study was the lack of multivariate analysis. This could have contributed to identify predisposing factor for health related quality of life impairment. Many of the aspects of quality of life were not evaluated such as the local health indicators, economic data, access and social exclusion, as well. Despite this fact, the current study adds several important findings to the existing literature about this subject. This is one of the few studies demonstrating the quality of life of pediatric patients with SCD compared to healthy children, especially in the social environment of Latin America countries. Our findings demonstrated that children and adolescents with SCD had lower scores of quality of life in the domains activities, social and school functioning when compared to healthy pediatric individuals.

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