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Original article

Socioeconomic and demographic characteristics of sickle cell disease patients from a low-income region of northeastern Brazil

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ABSTRACT

Objective: To characterize the socioeconomic and demographic aspects of sickle cell disease patients from the state of Rio Grande do Norte (RN), Northeast Brazil, and their adherence to the recommended treatment.

Methods: This cross-sectional descriptive study was performed at referral centers for the treatment of hematological diseases. One hundred and fifty-five unrelated individuals with sickle cell disease who went to these centers for outpatient visits were analyzed. All the patients, or their caregivers, were informed about the research procedures and objectives, and answered a standardized questionnaire.

Results: The patients were predominantly younger than 12 years old, self-declared as mulatto, lived in small towns fairly distant from the referral center, and had low education and socioeconomic levels. Individuals who were ten or younger were diagnosed at an earlier age. Almost 50% of the patients were taking hydroxyurea, 91.4% reported having received pneumococcal/meningococcal vaccinations and 76.1% received penicillin as antibiotic prophylaxis. However, the majority of them reported having difficulties following the recommendations of the physicians, mainly in respect to attaining the prescribed medications and transportation to the referral centers.

Conclusion: These individuals have a vulnerable socioeconomic situation that can lead to an aggravation of their general health and thus deserve special attention from the medical and psychosocial perspectives. Thus, it is necessary to improve public policies that provide Brazilian sickle cell disease patients with better access to medical treatment, living conditions, and integration into society.

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Introduction

Sickle cell disease (SCD) is one of the most common severe monogenic disorders worldwide.[1] The underlying molecular defect is a single nucleotide substitution (β^S – HBB; GAG>GTG; glu → val; rs334) in the gene that encodes the β-globin chain of hemoglobin. The resulting hemoglobin S (Hb S) polymerizes when deoxygenated, causing polymer-associated lesions of the red blood cells.[1,2] SCD includes several different genotypes including sickle cell anemia (Hb SS) and compound heterozygotes of Hb S with β-thalassemia (Hb S/β-thal) or with other types of hemoglobinopathies.[3]

The World Health Organization recognized SCD as a global public health problem, as the overall number of babies born with SCD between 2010 and 2050 is estimated at about 14.24 million.[4] Data from the Ministry of Health estimates that around 3500 children are born with sickle cell anemia each year in Brazil and the number of cases of the disease is between 25,000 and 30,000.[5]

The complications of this disease are numerous and can affect every organ and tissue in the body. The most common complications are pain crises, chronic anemia and its acute exacerbations, stroke, acute chest syndrome, infection, priapism, leg ulcerations, osteonecrosis, and cardiac and renal problems.[6] Complications can be acute, producing dramatic clinical findings, or chronic, disabling, and cause premature death.[2]

Specific phenotypic manifestations of the disease vary considerably in frequency and severity between patients and even in the same patient over time.[6] Both genetic and acquired factors contribute to this clinical variation. Among the acquired factors, the most important is the patient's socioeconomic conditions.[7]

Knowledge of the demographic and socioeconomic profile of SCD patients is essential to identify their needs, to contribute to improving resource allocation and to create and implement public health policies that benefit this population.[8] However, studies that address these aspects of the disease are relatively scarce in both the Brazilian and international literature.

Thus, this study aimed to characterize the demographic and socioeconomic aspects of SCD patients from the state of Rio Grande do Norte (RN), a socioeconomic vulnerable area of northeastern Brazil, and their adherence to the recommended treatment.

Methods

A cross-sectional descriptive study was performed at referral centers for the treatment of hematological diseases in RN: Hemocentro Dalton Cunha (Natal), Hospital Infantil Varela Santiago (Natal), and the Centro de Oncologia e Hematologia de Mossoró (Mossoró). The participants were unrelated SCD patients without cognitive impairment, who went to these centers from March 2011 to October 2013 for outpatient visits.

All the patients, or their caregivers, were informed about the research procedures and objectives, and those who agreed to participate in the study signed an informed consent form and answered a standardized questionnaire. When the patient was younger than 18 years old, it was answered by the caregiver.

The questions were orally asked by the interviewer without inducing responses. Questions aimed to collect medical history and the demographic and socioeconomic data of the patient, including age, ethnicity, ancestry, residence, schooling, employment situation, family income, age at diagnosis, use of hydroxyurea, prophylactic penicillin, immunization, and difficulties in following treatment, among others. Clinically relevant data were also taken directly from the patient's health records.

Data were collected in single individual interviews, and after collection they were input into a Microsoft Excel 2010 spreadsheet. Frequency distribution tables were used for the descriptive analysis of the categorical or nominal variables and the significance of differences between clinical characteristics by age group were estimated using the Chi-squared (χ^2) or Fisher exact test, as appropriate. The comparison of the age at diagnosis of SCD in age groups employed the Kruskal–Wallis analysis of variance (ANOVA) test, followed by multiple comparisons of mean ranks, using the Statistica software (version 7). Differences with a p-value ≤ 0.05 were considered statistically significant.

This study was conducted in accordance with the Helsinki Declaration as revised in 2008, and was approved by the Research Ethics Committee of the Universidade Federal do Rio Grande do Norte (UFRN, under protocol number 193/09) according to resolution 196/96 of the Conselho Nacional de Saúde, Brazil.

Results

One hundred and seventy-seven patients with clinical and laboratory diagnosis of SCD were interviewed. However, 22 were first- or second-degree relatives of other patients participating in the study and were therefore excluded from the analysis. Among the remaining 155 individuals, 109 (70.3%) had Hb SS, 23 (14.8%) were heterozygous for Hb S and β-thalassemia, 21 (13.5%) were heterozygous for Hb S and Hb C, and two (1.3%) presented the association between Hb S and hereditary persistence of fetal hemoglobin (HPFH).

The ages of the patients ranged from seven months to 48 years, with a median age of 12 years; the highest frequency of individuals was in the age group ≤5 years (29.0%), followed by the 11- to 15-year-old group (18.7%). The majority of the individuals were male (52.9%), and self-declared as mulatto (65.8%), but with no information about their ethnic ancestry (83.9%). However, indigenous ancestry was predominant (12.3%) among patients who informed their ancestry. A high percentage of the patients (43.2%) lived in small towns, at least 60 km away from the referral centers (Table 1).

Of the over 18-year-old patients, 51.9% had not completed high school, 50.0% were unemployed, and 19.2% were retired or receiving social security benefits. Most of the caregivers of the under 18-year-old patients only completed primary school education and the majority (55.3%) were working in regular jobs. Most of the patients (52.7%) had a household income of up to one minimum wage in Brazil, about US$ 240.00, and one third (34.2%) did not receive any social benefits from