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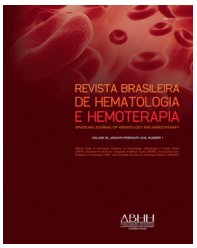

Original article

Clinical and laboratory profile of patients with sickle cell anemia

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ABSTRACT

Objective: This study aimed to describe and analyze clinical and laboratory characteristics of patients with sickle cell anemia treated at the Hemominas Foundation, in Divinópolis, Brazil. Furthermore, this study aimed to compare the clinical and laboratory outcomes of the group of patients treated with hydroxyurea with those patients that were not treated with hydroxyurea.

Methods: Clinical and laboratorial data were obtained by analyzing medical records of patients with sickle cell anemia.

Results: Data from the medical records of 50 patients were analyzed. Most of the patients were female (56%), aged between 20 and 29 years old. Infections, transfusions, cholecystectomy, splenectomy and systemic arterial hypertension were the most frequent clinical adverse events of the patients. The most frequent cause of hospitalization was painful crisis. The majority of patients had reduced values of hemoglobin and hematocrit (8.55 ± 1.33 g/dL and 25.7 ± 4.4%, respectively) and increased fetal hemoglobin levels (12 ± 7%). None of the clinical variables was statistically significant on comparing the two groups of patients. Among hematological variables only hemoglobin and hematocrit levels were statistically different between patients treated with hydroxyurea and untreated patients (p-value = 0.005 and p-value = 0.001, respectively).

Conclusion: Sickle cell anemia requires treatment and follow-up by a multiprofessional team. A current therapeutic option is hydroxyurea. This drug reduces complications and improves laboratorial parameters of patients. In this study, the use of the drug increased the hemoglobin and hematocrit levels of patients.

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Introduction

Sickle cell anemia (SCA) is an inherited autosomal recessive disease characterized by the presence of homozygous hemoglobin S (Hb S). It is caused by a single nucleotide mutation that substitutes glutamic acid for valine in the sixth position of the β-globin gene.[1] During hypoxic conditions, the red blood cell becomes sickled and the resulting change in structure restricts circulation causing obstruction of the blood flow within the capillaries and early destruction of the cell.[2]

Clinical manifestations of SCA vary from mild, that is, almost asymptomatic, to severe forms that are associated with high mortality rates.[3] Clinical manifestations usually appear after three months of age, when the concentration of fetal hemoglobin (Hb F) decreases.[2] Most systems are liable to vaso-occlusive processes possibly resulting in multisystem failure.[4–6]

A definitive cure is not currently available for patients with SCA. Existing therapies are only focused on symptom management and do not alter the natural history of the disease. These therapies are comprised of hydration, prevention of infections, pain management, proper nutrition and precautions against adverse weather conditions. Thus, additional therapies are needed to prevent complications without subjecting patients to the increased morbidity and mortality associated with highly aggressive approaches such as hematopoietic stem cell transplantation (HSCT).

Currently, hydroxyurea (HU) is the only medical modality with proven efficacy in patients with frequent symptoms related to SCA.[7,8] HU is known to increase Hb F levels, improve hemoglobin concentrations and mean corpuscular volume, and reduce the number of reticulocytes. Another favorable response of treatment is that it does not only reduce the expression of adhesion molecules, but also decreases the number of receptor proteins located on endothelial cells. Therefore, HU decreases vascular adhesion which contributes by diminishing the number of vaso-occlusive crises.[9,10]

SCA is an inherited disease with high prevalence and mortality rates.[11] However, the literature is scarce on local epidemiological studies in Brazil. Hence, this study aims to analyze the clinical and laboratorial characteristics of patients with SCA who live in the macro region of Divinópolis, Minas Gerais treated in the Hemominas Foundation. Furthermore, this study aims to compare the clinical and laboratorial outcomes in two groups of patients; those treated with HU to those that are not treated with HU.

Methods

Study sample and data collection procedure

A retrospective study was carried out based on information extracted from medical records. This study was conducted at the Hemominas Foundation, a blood center in Divinópolis, Brazil. All patients from that macro region diagnosed with SCA (limited to the homozygous Hb SS genotype) and followed-up from August 2012 to August 2014 were included. Initially the planned sample was composed of 57 patients that is, all cases of SCA at the Hemominas Foundation. However, seven patients were excluded from analysis (five were not followed-up in the previous two years, one patient died and one patient was transferred to another blood center).

This study was approved by the Research Ethics Committee of the Universidade Federal de São João Del Rei, Campus Centro Oeste (# 477.473) and by the Research Ethics Committee of the Hemominas Foundation (# 506.674).

A standardized data extraction form was used to collect information from patients' medical records. All data collected refer to the period of interest (August 2012 to August 2014). The form contained the following information: age, date of diagnosis, date of starting treatment, adherence to the institutional vaccination protocol (protection against influenza, meningococcus and pneumococcus), clinical and therapeutic aspects, and laboratory tests results. The arithmetic mean was calculated from the last five laboratory test results within the period of the trial.

Statistical analysis

Descriptive statistics are used to report the variables of interest. Categorical variables are reported as absolute and relative frequencies. Continuous variables are presented as means and standard deviation or median and interquartile range if the variable showed non-parametric distribution. Distribution of the data was tested using the Shapiro–Wilk test. Normally distributed data were analyzed using the Student's *t*-test, whilst non-parametric data were analyzed using the Mann–Whitney *U* test. Pearson's chi-square test or Fisher's exact test, as appropriate, were used to associate exposure with outcomes. These analyses were performed using the Statistical Package for Social Sciences (SPSS Inc., Chicago) version 22 for Windows. The level of significance was set at 5% (p-value <0.05).

Results

Data from the medical records of 50 patients were analyzed. Females, with a total of 28 (56%), were predominant in the sample. The age range of the patients was 2–54 years with a mean of 25.4 ± 12.9 years. When grouped by age, the majority of the patients were between 20 and 29 years (36%) while the over 50-year-old age group was the smallest (4%).

The age at diagnosis was reported in 30 patient records, with 14 (46.7%) patients diagnosed at less than one year of age (median: 1.0; interquartile range: 0.0–10.5 years) as part of the Newborn Screening Program in Minas Gerais. Regarding the number of medical appointments over the previous two years, 50% of patients had 17 medical appointments or more (interquartile range: 11–32 consultations).

Clinical characteristics

Seven patients (14%) underwent splenectomy and 15 patients (30%) were submitted to cholecystectomy. Nine of the 50 patients had systemic arterial hypertension (18%). Splenic sequestration was not mentioned in any of the records within the period studied. The vaccination protocol was described in 31 records with 24 patients (77.4%) adhering to the protocol established by the Brazilian Health Ministry[12]; all under

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