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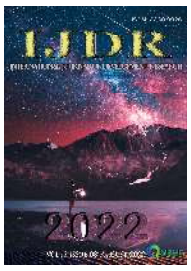

RESEARCH ARTICLE OPEN ACCESS

EVALUATION OF CLINICAL AND LABORATORY ASPECTS IN ADOLESCENTS WITH SICKLE CELL ANEMIA, BEFORE AND AFTER TREATMENT WITH HYDROXYUREA

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ABSTRACT

Objective: To evaluate clinical and laboratory aspects of adolescents with Sickle Cell Disease treated with hydroxyurea. **Methodology:** a retrospective cohort study of patients with sickle cell anemia. The population will be adolescents who have been on hydroxyurea treatment for at least 2 years. Patients with other hemoglobinopathies, who abandoned treatment or died will be excluded. Data will be collected from medical records from September to October 2022. All ethical and legal principles will be respected and a significance level of 5% ($p<0.05$) will be considered.

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INTRODUCTION

The term sickle cell disease (SCD) refers to a group of conditions with a predominance within the erythrocytes of a variant hemoglobin called Hb S (Naoum, P.; Naoum, F, 2004). Hb S can occur as homozygous (HbSS or sickle cell anemia), heterozygous (HbAS or sickle cell trait), in double heterozygosity (HbSC, HbSD), associated with Hereditary Persistence of fetal Hb (S/PHHF) or in association with thalassemias. : S-alpha thalassemias (Sα+thal or Sα0thal) and S beta thalassemias (Sβ+thal or Sβ0thal) (Naoum, P.; Naoum, F, 2004). In Brazil, FD is heterogeneously distributed in regions according to the presence of the African population "uprooted from their countries to slave labor", and is part of the group of relevant diseases and conditions that affect the Afro-descendant population (Cançado e Jesus, 2007). It is estimated that approximately 4% of the population and 6% to 10% of Afro-descendants have Hb AS, and 20 to 30 thousand Brazilians have DF (Araújo, 2015). Sickle cell disease is caused by a mutation in the gene that produces hemoglobin A, resulting in another altered hemoglobin called S (Galiza Neto; Pitombeira, 2003). The underlying change is due to a single nucleotide substitution (GTG for GAG) in the gene encoding the α-globin chain (Silva-Pinto *et al*. 2013). Hb S is a protein that, under deoxygenation conditions, has the unique property of polymerizing into long fibers, reducing the deformability of erythrocytes (Silva-Pinto *et al*. 2013). HbS becomes insoluble, changing the erythrocyte shape, biconcave disk, to a structure that resembles a sickle, called erythrosickling and/or sickling (Naoum, Naoum 2004; Zago; Pinto, 2007). In this way, it hampers erythrocyte rheology and survival, manifesting in hemolytic anemia and microvascular vaso-occlusion cycles that can lead to target organ ischemia-reperfusion injury and infarction (Sundd *et al*., 2019). Regarding the treatment with Hydroxyurea (HU), a double-blind, randomized, multicenter clinical trial, carried out with 299 adult patients with sickle cell anemia, treated with HU, obtained in the results, reduction of hospitalizations, annual rates of crises, chest syndrome and transfusions compared to to patients who received placebo (Charache *et al*., 1995; Belini Junior *et al*, 2015); Another study that followed adult patients with SCA for 17.5 years to examine the risks and benefits of HU, the authors suggest that the use of HU in the long term is safe and may decrease mortality (Steinberg *et al*, 2010). With the establishment of the diagnosis of SCD, patients need follow-up at the SUS Services. It should be noted that the guidelines of the National Comprehensive Care Policy aim to "ensure the follow-up (by the SUS) of people with hemoglobinopathies, diagnosed by the PNTN, by a multidisciplinary team" (Brasil, 2010) and there are gaps in research that explore this theme in Brazil. A study of professionals who work at the entrance door of Health Services carried out in Minas Gerais, showed the need for permanent training of these professionals in DF, to make a difference, and depending on the situation, the attitude of this professional can be decisive in the care of the patient with sickle cell disease (Kikuchi *et*

al., 2018). From the perspective of Comprehensive Care for the population with SCD, it is intended to advance the discussion, specifically for adolescents with SCD. A mortality study of patients with sickle cell anemia of different age groups, carried out in Mato Grosso do Sul, showed a low survival rate in SCA at 18.88 years (Pompeo *et al*, 2021). This evidenced epidemiological indicator, added to the transition phase for adults, deserves to be investigated. Thus, this study intends to seek answers to the question: what are the effects of hydroxyurea on clinical and laboratory aspects in adolescents with sickle cell anemia treated in Mato Grosso do Sul.

METHODS

Type of study: This is a cohort study, with retrospective data collection from medical records of adolescent patients diagnosed with sickle cell anemia.

Location and Period of Research: The research will be carried out at the Hospital Regional de Mato Grosso do Sul (HRMS), located in the city of Campo Grande/MS, a reference hospital for the care of patients with Sickle Cell Anemia in the State of Mato Grosso do Sul (MS). Data will be collected from the records of the Medical and Statistical Archive Service from September 2022 to October 2022. Records of adolescents with records from 2002 to 2011 will be analyzed. This period was chosen to limit the age of adolescent patients (aged 10 to 19 years) (BRASIL, 2007).

Population and sample: The population will be composed of medical records of adolescent patients with a confirmed medical diagnosis of sickle cell anemia. The sample will be a census, composed of the medical records of patients who meet the inclusion criteria. For this study, adolescence will be considered the WHO definition (Brasil, 2007), people aged 10 to 19 years. To evaluate the efficacy and toxicity of HU, at least two years of treatment are required (Brasil, 2018; Brasil, 2010). The period before corresponds to two years prior to the date of the first medical prescription of hydroxyurea, and the period after, the four years following the start of treatment.

Inclusion criteria for participants: It is estimated that there are approximately 60 adolescents in the HRMS being followed up with HU.

The medical records of patients who meet the following criteria will be included the Diagnosis, according to the International Classification of Diseases (ICD-10):

- ICD D57. 0 Sickle cell anemia with crisis;
- ICD D57.1 Sickle cell anemia without crisis
- Patients of both sexes;
- Age greater than or equal to 10 years and less than 19 years;
- Patients on hydroxyurea treatment for at least two years.

Exclusion criteria for participants

Medical records of patients who present:

- Diagnoses of other hemoglobinopathies;
- Patients who are not being followed up due to abandonment and death;
- Patients under 10 years of age or over 19 years of age.
- Patients with less than two years of hydroxyurea treatment.

Data collect: Data will be collected after project approval by CEP/UFMS. The collection of research data will be carried out at the Medical Records Service of HRMS, in Campo Grande - MS, following the institution's protocols. A data collection instrument based on the research variables will be used. Identification variables (Gender, date of birth, municipality of birth and municipality of residence); clinical variables (age at initiation of treatment with HU, date of initiation of the first medical prescription for the treatment of HU, dosage (mg) of capsules (at all times noted in the medical record), patient weight for calculation of hydroxyurea dosage, transfusions blood pressure, complications of AF); hospitalization variables (frequency, days of stay and reason for hospitalization); laboratory variables (erythrocytes/Red cells (He M/µL), hematocrit (Htc %), hemoglobin (Hb g/dL), reticulocytes (%), mean corpuscular hemoglobin concentration (HCCM g/dL), mean corpuscular hemoglobin (MCM pg) , mean corpuscular volume (MCV fL), leukocytes (% /µL), platelets (Plaq µL), neutrophils Abso %, fetal hemoglobin – Hb F, ferritin, transferrin saturation, creatinine, TGO/AST, TGP/ALT, serum iron and free iron concentration.

Statistical analysis of data: The data collected will be entered into a Microsoft Office Excel spreadsheet. To minimize errors in data entry, double typing will be performed and the error checking will be analyzed in the statistical program SPSS version 20.0. Data will be analyzed comparing two moments, before and after treatment with HU, and will occur in the same patient. After analyzing the data, the information will be presented through figures and tables. The level of significance of the data will be verified through specific tests and considered a level of significance of 5% ($p < 0.05$), using the statistical program SPSS version 20.0.

RESULTS AND DISCUSSION

By proposing the present study, it is expected to promote the reflective process of health professionals and society regarding the clinical, social and cultural issues relevant to sickle cell anemia in adolescents. It is believed that carrying out this study, a pioneer in the state, can sensitize managers about the improvement of the necessary structure for the care of adolescents with SCA and bring subsidies to professionals and health institutions to improve the organization and planning of the processes related to the treatment. with HU. There are no specific criteria on what to describe results on treatment with hydroxyurea (HU) and other drugs available for a specific knowledge of adolescents with inflammatory diseases. Due to an existing review on the topic related in the literature, we consider it opportune to carry out a scope to answer the question: what drugs are available to adolescents with sickle cell disease whose pathophysiological mechanism for severe acute crises.

FINAL CONSIDERATIONS

The present study presents a topic of worldwide relevance. The development of this research is within the elaborate schedule, and at the end with the results obtained, they will be communicated in future publications and it is expected to contribute to interventions based on scientific evidence, as well as to add to the clinical reasoning used in the treatment of adolescents with sickle cell anemia

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