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Mortality in sickle cell anemia: a retrospective cohort study

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ABSTRACT

Objective: To epidemiologically analyze the mortality of the patients with sickle cell anemia in two reference centers for treatment in the state of Mato Grosso do Sul. **Method:** An epidemiological study with a quantitative approach and of the descriptive and analytical type, by means of a retrospective, individualized cohort design. It will be carried out in two public hospitals of reference for the treatment of Sickle Cell Anemia (SCA) in Mato Grosso do Sul and will use as a unit of analysis individuals diagnosed with SCA followed-up in these services from 1980 to 2018. **Expected results:** It is expected to historically describe the course of SCA in order to obtain subsidies to verify the outcomes, among them the deaths and their respective causes, to enable strategies for treatment and control of the disease and its complications.

Keywords: Chronic disease; Sickle cell anemia; Mortality; Survival; Epidemiology.

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INTRODUCTION

Sickle Cell Disease (SCD) is a hereditary hematological disease, which results from changes in the hemoglobin subunit beta. Among the main forms of SCD is Sickle Cell Anemia (SCA), which consists of the homozygous state of the disease, considered to be of higher prevalence and severity when compared to the others[1].

After the implementation of neonatal screening programs, the use of Hydroxyurea, penicillin and the immunization with special immunobiologicals, improvements were obtained in the survival of individuals with SCA; however, the mean survival has not yet surpassed the fifth decade of life, estimated to be around 30 years of age on average[1,2].

It is estimated that SCD, especially the homozygous form, is responsible for at least 5% to 16% of all the mortality in children under five years old in Africa[3]. In this context, mortality and low survival of patients with SCA are conditions that require increased efforts by the scientific community and public policy managers. However, the world literature still proves to be insufficient to promote changes in the global panorama on the life expectancy of these patients.

Thus, this research is justified by the scarcity of studies on the subject and by the lack of publications that comprehensively address all the aspects involved with the survival and mortality of patients with SCA. In addition, this study will support researchers, professionals, and health managers regarding the next steps to be taken in relation to the treatment of SCA.

OBJECTIVES

To epidemiologically analyze the mortality of the patients with sickle cell anemia in two reference centers for treatment in the state of Mato Grosso do Sul.

METHOD

An epidemiological study with a quantitative approach and of the descriptive and analytical type, by means of a retrospective, individualized cohort design.

Variables such as the use of hydroxyurea and its association with clinical and laboratory data, number of blood transfusions, age, number of hospitalizations, presence of complications such as pain and occlusive vessel crises, infections, and splenic sequestration, among others, will be evaluated. In addition to mortality data, the focus of this study.

It will be carried out in two public institutions of reference for the treatment of SCA in the state of Mato Grosso do Sul (MS). The population will be composed of individuals with SCA followed-up by the analyzed services, by means of a census sample.

Data from all the medical records of the individuals with a medical diagnosis of SCA confirmed by the laboratory and followed-up by the two listed hospitals will be included. Those who do not have sufficient data to meet the objectives of the study, as well as those that are illegible or lost, will be excluded.

The data of the patients followed-up between 1980 and 2018 will be analyzed. This time frame is justified by the occurrence of the first care measures of reference for SCA carried out in the institutions listed in the late 1970s and early 1980s. In addition, the choice of 2018 as the end time was so that the study

patient had at least one year of follow-up for the disease. Data collection will take place in the second half of 2019 with completion expected by the end of the same year.

The data will be obtained from secondary sources. The cases will be identified by tracking them in the database of the Research, Teaching and Diagnosis Institute of the Association of Parents and Friends of the Exceptional (*Instituto de Pesquisa, Ensino e Diagnóstico da Associação de Pais e Amigos dos Excepcionais*, IPED/APAE) of Campo Grande/MS, the body responsible for the diagnosis of hemoglobinopathies in the state. To obtain the information, a hospital record will be used with the application of a data collection instrument designed specifically for this study. The data will be encoded, categorized and stored in a computerized database and computed using the SPSS (*Statistical Package for the Social Sciences*) statistical package, version 24.0 for Windows, with a significance level of 0.05.

The Chi-square test or Fisher's Exact test will be applied for the categorical variables, according to the nature of each variable. For the non-categorical categories, the T-Student and Mann Whitney tests will be used. The risk factors and predictors of mortality will be evaluated by the Cox regression model and the Krushall Wallis test will be applied to compare individuals who died with those who survived. The survival analysis will be done using the Kaplan-Meier method.

This study will meet all the regulatory guidelines for research involving human beings and was approved by the Ethics and Research Committee on Human Beings of the Federal University of Mato Grosso do Sul (*Universidade Federal de Mato Grosso do Sul*, UFMS) in ethical and methodological aspects, protocol No. 3,226,971 and CAAE No. 07575318.7.0000.0021.

EXPECTED RESULTS

It is expected to describe the historical course of SCA in each individual followed-up by the institutions described, in order to obtain subsidies to verify the outcomes, among them the deaths and their respective causes.

In addition, this study proposes to produce benefits for the patients with SCA, as well as for the health services, professionals, and public policy managers. When identifying what affects the health of this population, to what extent this occurs, as well as the generation of limitations or disabilities in the individuals, the description for the main causes of death is expected to outline strategies for treatment and control of the disease and its complications, as well as to identify factors that may increase the survival of this specific clientele.

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