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ORIGINAL ARTICLE

Profile of pediatric patients with sickle cell disease admitted to Children's Hospital of the Irma Dulce Social Works in Salvador (BA)

Ariadne Carvalho Godinho[1], Ivana Paula Ribeiro Leite[1]

Headings:
Epidemiology,
Anemia,
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Pediatrics.

Abstract

Objectives: To describe the profile of patients with sickle cell disease admitted to the Childrens Hospital of the Irma Dulce Social Works in Salvador city, Brazil. **Methods:** Descriptive study through the analysis of hospitalization data registered in the book of Medical Residence in Pediatric Hematology and in the Hospital Information System, from March 2018 to March 2019. **Results:** The main one cause of hospitalization is due to occlusive vessel crisis (59.5%) and the age group most affected is from 01 to 05 years (30%). Patients from Salvador city, capital of Bahia state, account for 65%. The average hospital stay is six days. Most patients still do not use hydroxyurea and 6% do not undergo regular outpatient follow-up. **Conclusions:** Knowing the profile of patients with sickle cell disease can serve as a basis for improving the specific care that this public needs and for the development of actions for the early detection of the disease and the implementation of reference centers in the interior of the state of Bahia.

[1]Health Department of the State of Bahia, Medical Residency in Pediatric Hematology and Hemotherapy - Salvador - BAHIA - Brazil.

Correspondence to:
Ariadne Carvalho Godinho.
Health Department of the State of Bahia. 4th Avenida Centro Administrativo da Bahia, 400, Centro Administrativo da Bahia, Salvador, BA, Brazil.
CEP: 40301-110. E-mail: ariadnegodinho@gmail.com

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INTRODUCTION

Sickle Cell Disease (SCD) is one of the most common diseases in the world. It is a set of chronic and hereditary hematological pathologies that have in common the presence of hemoglobin S (HbS) in red blood cells. HbS, when deoxygenated and in high concentration, presents a change in its conformation and assumes the shape of a sickle, generating the vaso-occlusive phenomenon, which causes acute and chronic pain, the main clinical characteristic of SCD and the main reason for hospitalization[1].

The hemoglobin S gene is of high frequency in Brazil, being more common in the Southeast and Northeast regions. It is estimated that three thousand children a year are born with this disease in the country[2] with a tendency to reach an increasingly significant portion of the population due to the high degree of miscegenation[3]. For this reason, sickle cell disease can be considered a public health problem in the country.

It is a hereditary disease with a chronic course, which causes a great impact on the entire family and, therefore, all aspects that involve it need to be well known by the health services[4]. A chronic disease is a source of permanent stressors, which can generate difficulties in family relationships, interaction with colleagues, academic performance and the development of a positive self-image[5].

The present study has the main objective of tracing the profile of hospitalizations of patients with SCD at the Hospital da Criança das Obras Sociais Irmã Dulce (HC-OSID) in Salvador (BA), so that the results can serve as a basis for improving care. specific needs that this public needs and for the elaboration of measures for the early detection of the disease and the implementation of reference centers in the interior of the state.

METHODOLOGY

The study is descriptive and was carried out through the analysis of data from hospitalizations of patients with SCD registered in the notebook of the Medical Residency in Pediatric Hematology and in the HC-OSID Medical Records Information System. The time frame was from March 2018 to March 2019, with no separation regarding the specific genotypes of hemoglobinopathies. The prioritized information was gender, age group, reason and length of stay, outpatient follow-up, patients' place of residence, use of hydroxyurea.

RESULTS AND DISCUSSION

The total number of hospitalizations during the study period was 90, 52% of which were female patients. The most frequent age group was between 1 and 5 years old (30%), followed by the group from 5 to 10 years old (28%), as shown in Figure 1. A similar result regarding the age group of pediatric hospitalizations for SCD was found in a study[6] carried out in Espírito Santo in 2016. Of the patients surveyed, 17% had an average of three readmissions within a year. The average

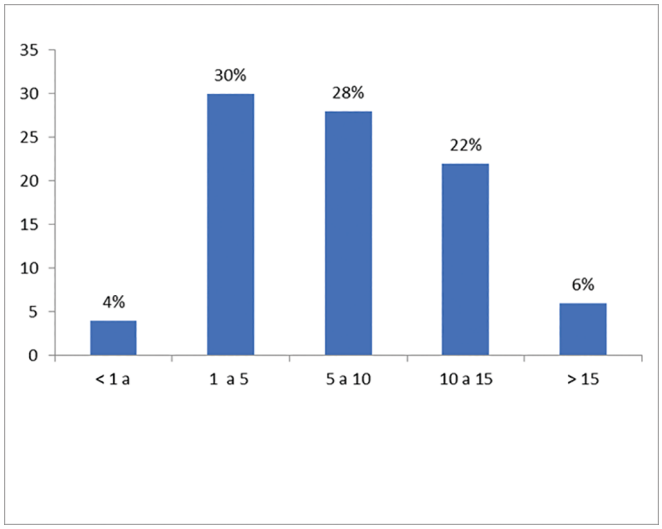


Figure 1. Percentage of patients by age group.

period of hospitalization was six days, with a minimum stay of three and a maximum of 16 days.

The causes of hospitalization were Vaso-Occlusive Crisis (59.5%), Acute Thoracic Syndrome (19.5%), Fever without a defined focus (11%), Splenic Sequestration (8%) and Post-operative Cholecystectomy (2%). Pain crises caused by body microcirculation obstruction are one of the main symptoms of sickle cell disease, being responsible for most hospitalizations and emergency care[7]. Among the group studied, only 18% used hydroxyurea medication at an average dose of 18.6mg/kg. In another study also carried out in the city of Salvador, it was found that only 31% of patients with SCD used this medication[8]. Hydroxyurea is a medication suitable for SCD, which has as one of its forms of action the increase in fetal hemoglobin and, consequently, a decrease in the acute events of the disease. Thus, it is desirable that this drug be prescribed to a greater number of SCD patients. However, for this to occur, regular monitoring of the patient with a hematologist is necessary.

Hematopoietic stem cells, indicated for children who have a compatible donor in the family[1] and who have severe cases, such as the occurrence of stroke. However, crizanlizumab, a medication that acts on the downregulation of P - selectin in endothelial cells and platelets, may contribute to cell-cell interactions that are involved in the pathogenesis of vaso-occlusion[9], promising to minimize crises of sickle cell pain and, thus, improve the quality of life of patients with SCD. However, no patient was found in the study who used this drug.

Regarding the follow-up of patients with SCD, 23% do so at HEMOBA, as shown in Figure 2. What stands out is that 6% of patients do not have regular follow-up, most likely due to the distance from their home town to the capital of Bahia, there is given that few cities in the interior of the state have specialized assistance in pediatric hematology.

Of the group studied, 65% are from Salvador, as illustrated in Figure 3. Despite being a minority, patients from the interior of the state add up to a significant number

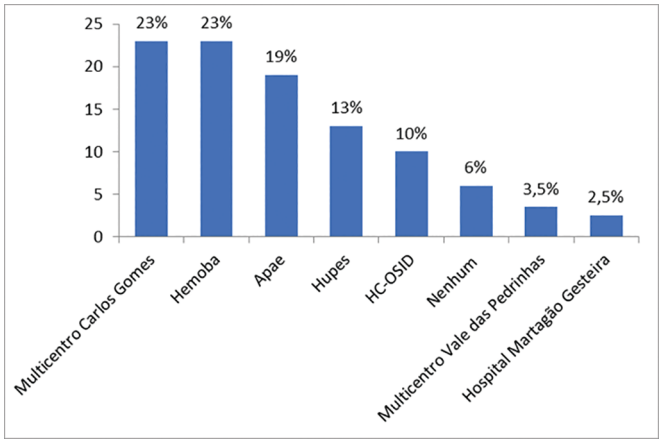


Figure 2. Health Units where patients undergo outpatient follow-up.

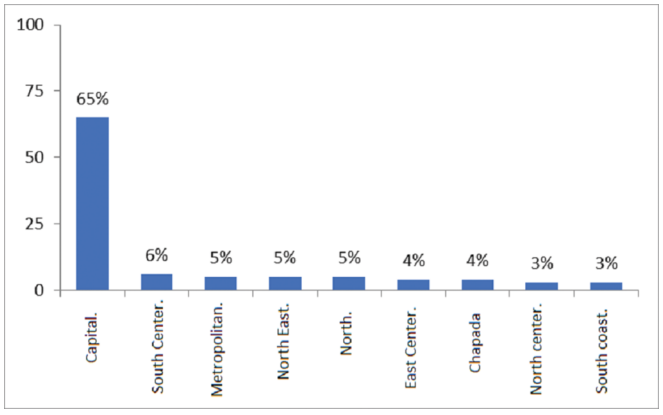


Figure 3. Regions of Bahia where the patients came from.

and, therefore, ends up generating the debate about the decentralization of specialized care in pediatric hematology. In this context, specialized care in FD should also be offered in other regions of the state, aiming at better patient care, in view of the inconvenience (logistic and financial, among others) of families traveling long distances to receive adequate care.

Public health policies need to move towards universal assistance that includes, among other things, greater adherence to treatment. The care demands related to the scheduling of specialized consultations, high-cost exams, hospital admissions and elective surgeries are not yet structured/regulated in the health care network, which significantly affects the comprehensive care of these patients[4].

The appropriate approach to the patient with sickle cell disease depends on the performance of a multidisciplinary team, on the participation of the family and the community, always aiming at the integrality of health actions for qualified assistance[5]. Thus, it is clear the need for patients with SCD to be attended by a team of professionals specialized in the city or region in which they live, even for a more adequate assistance.

CONCLUSION

The profile of pediatric patients hospitalized for sickle cell disease presented in this study has the potential to assist in the development of health care measures for patients with the disease. The main cause of hospitalization is a vaso-occlusive crisis, and few patients use hydroxyurea , a drug that reduces the acute events of SCD. There is a significant number of patients from the interior of Bahia. Therefore, their regular follow-up with pediatric hematologists in the city or region where they live should be prioritized, aiming at a more adequate and integrated care. For this, it is essential to implement reference centers also in the interior of the state for early diagnosis, reduction of morbidity, hospitalizations and costs associated with clinical follow-up, and, mainly, improvement in survival and quality of life of patients. children with SCD, especially those under five years of age.

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