Emergency care necessity for sickle cell disease patients at Rio de Janeiro State Coordinating Blood Bank

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Hemoglobinopathies, in particular sickle cell disease, is the most prevalent group of genetically transmitted diseases in the Brazilian population and should thus be treated as a public health problem. Many of these patients frequently present with complications and require emergency care at the blood bank Coordinator in Rio de Janeiro. This study was developed with the aim of characterizing the emergency assistance required by sickle cell disease patients registered in the blood bank from January 2007 to December 2008. A retrospective study of medical records was made of 78, mostly children, patients from the date of their registration until December 2009. Most attendances (63.7%) were not considered emergency care. The use of specialized services for cases that do not require this level of complexity may saturate the capacity of these centers. However, delay of intervention for complications due to the transportation of patients to specialist centers may lead to deterioration in the clinical condition.

Keywords: Anemia, sickle cell; Emergency medical services; Hospital Care; Retrospective studies

Introduction

In Brazil, due to miscegenation, sickle cell disease is the most prevalent genetic disease and is emerging as a major public health problem, that should be addressed in an intersectoral and integrated policy. Despite this epidemiological relevance and the commitment of some professionals and health policies in Rio de Janeiro to decentralize care of these patients, the vast majority of patients are still only treated in large referral centers. The development of the hematology support network in Rio de Janeiro State (Hemorrede) is a political, social and economic challenge in that it aims to provide better accessibility to care, minimizing the physical and emotional stress, lowering the cost of care and stimulating the growth of this network beyond the walls of specialized units.

The biggest challenge, however, is to demystify sickle cell disease as a "disease for experts", i.e., a disease that only hematologists are able to treat. With the success of the neonatal screening program, pediatricians have an important role in monitoring newborn babies.

In addition to primary care services it is necessary to take a careful look at early intervention in clinical complications. The time required to transport a patient to a specialist center may be crucial for the outcome. Thus, physicians and other healthcare professionals should be able to provide adequate care to sickle cell disease patients with clinical complications.

From the perspective of supporting training programs and training healthcare teams to help to demystify and reduce the stigmatization of sickle cell disease and to serve as a basis in the organization of healthcare services, this work was developed with the aim of characterizing the profile of emergency care of sickle cell disease patients registered in the Rio de Janeiro State Coordinating Blood Bank from January 2007 to December 2008.

Methods

The study was conducted in accordance with Resolution CNS 169/9613 and approved by the Research Ethics Committee (CEP HEMORIO No 177/09). After characterizing the sociodemographic profile of sickle cell disease patients registered in the Rio de Janeiro State Coordinating Blood Bank in the period of 2002-2008, a retrospective study of the clinical profile of emergency treatment of these patients was performed.

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For this, all sickle cell disease patients (n = 307) admitted to the blood bank from January 2007 to December 2008 were selected. To analyze sociodemographic data and care needs, 78 patients were enrolled in this study (44 from 2007 and 34 from 2008) by random sampling using the Epidat® software version 3.1. The medical records of these patients were analyzed to characterize the number of consultations and the type of care provided by the Blood Bank Emergency Department until December of 2009 including: diagnosis, evolution and outcome. Data of up to 24 months for patients enrolled in 2007 and up to 12 months for patients enrolled in 2008 were stored in a database created in the EPIINFO® statistics program.

An adaptation of the Manchester Triage System(15) was used to characterize the severity of complications and the emergency care provided focusing on the sickle cell disease and related lesions with the aim of objectively and systematically identifying criteria of severity that indicate the clinical priority with which patients should be attended.

Results

Table 2 shows that nearly 30.0% of care provided in the Emergency Department was not, in reality, emergency assistance. In these cases the patients should have been seen in an outpatient care facility, as patients went to the Emergency Room to obtain the results of laboratory tests and to renew prescriptions of medications normally or frequently taken by the patient because they missed a scheduled outpatient visit, among other reasons. A total of 34.8% of cases could have been taken care of in any other primary care clinic or outpatient service near to the residence of the patient because the complaint was not characterized as hematology/hemotherapy specialized care. Only 12.6% of complications were severe requiring immediate emergency care, as they presented serious harm related to the underlying disease, such as pneumonia, spleen sequestration and painful crisis.

Table 3 presents the most common health complaints of patients and their caregivers that led to visits to the Coordinating Blood Bank Emergency Department. The most common complaints were fever (35.8%), pain (15.1%) and vomiting/diarrhea (10.1%).

Table 4 shows the main clinical diagnoses of these patients in the emergency room. Almost one quarter (23.7%) of patients had no medical complication noted during the interview and clinical examination that justified the need for care in the emergency room. A further 20.2% had symptoms related to upper respiratory tract infection (tonsillitis, pharyngitis, and rhinitis) and 9.1% had spleen sequestration of varying severity ranging from slight swelling of the spleen with no clinical implications to conditions that required emergency transfusions of packed red blood cells. Despite the complaints of pain listed in Table 3, only 7.1% of patients were diagnosed with painful crisis. At the time of care many patients did not need intravenous medications but had their complaints managed with only oral analgesia. The item "other
complaints” included all complaints that were reported less than five times such as conjunctivitis, constipation, exanthematous diseases, hypertension, impetigo and epigastric pain.

Table 5 shows that in 92.4% of emergency visits, the patient was discharged from the Emergency Department. Only 7.1% were hospitalized. In this study period one patient (0.5%) died during emergency care.

The times of stay in the emergency room are shown in Table 6. About 48% of patients remained for up to one hour, 5.5% remained for more than 48 hours with the median time being two hours for all patients.

Of the 198 emergency calls, only 48 (24.2%) required transfusional support with transfusions of packed red blood cells. However, with the exception of the 18 cases (9.1%) of splenic sequestration requiring urgent transfusion therapy, within 3 hours of the transfusion request by the attending physician, all the cases were considered non-urgent transfusions. There were no reports of transfusions of extreme urgency (immediate infusion without checking compatibility).(16)

### Discussion

Early diagnosis of sickle cell disease is extremely important to reduce the morbidity and mortality of those affected by adopting effective measures to reduce the phenomena that lead to hemolysis and vascular occlusion so common in sickle cell disease.(1,11,17)

The reduction in mortality in sickle cell disease and the increase in life expectancy of these individuals augments the need and demand for healthcare services of patients with hemoglobinopathies.
The Coordinating Blood Bank will be unable to absorb all the demand for care and will require support from other health facilities distributed across the state. A better solution of cases by primary healthcare units will reduce the demand for specialized consultations and examinations directing of cases by primary healthcare units will reduce the demand health facilities distributed across the state. A better solution all the demand for care and will require support from other levels of the healthcare system, the ideal is that this first visit, the initial guidance and early prophylactic and therapeutic treatment are carried out by multidisciplinary teams of primary healthcare clinics and of the Family Health Program as, although incurable, when sickle cell disease is diagnosed and treated promptly, it can be controlled by general measures that prevent complications, thereby significantly reducing morbidity and mortality. Thus wide-ranging educational programs are needed for professionals in the Family Health Program.

Secondary and tertiary care institutions, which serve as referral centers for the primary healthcare clinics, should also be prepared to absorb this demand for treatment of clinical complications that do not require a hematologist. This professional, the hematologist, should then be able to concentrate on monitoring the most serious and complicated cases, as well as coordinating implementation of a social support network for sickle cell disease patients.

Conclusion

It was concluded that most of the clinical complications of sickle cell disease patients treated at the Emergency Unit of the Rio de Janeiro Coordinating Blood Bank were not emergency situations. Clinical situations that were classified as emergencies due to the underlying disease should not necessarily be treated in the Coordinating Blood Bank as many complications can be solved in emergencies departments of general hospitals.

From the data presented, some questions are raised: why do patients not use emergency services closer to their homes? Why are simple cases that could be treated in primary healthcare clinics referred to the Coordinating Blood Bank?

The Coordinating Blood Bank must be seen as a referral facility for more serious cases and technical guidance and should not be solely responsible for the direct care of these patients. Another interesting aspect to be highlighted is the necessity to include the National Family Health Program in the care of these patients and their families; much of the
work of the Family Health Program is focused on health education, disease prevention and early identification of complications.

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