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.[1-9] 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.[10] With the success of the neonatal screening program,[11,12] pediatricians have an important role in monitoring newborn babies.

In addition to primary care services it is necessary a 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[13] 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.[14]

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Table 1 - Classification of severity and emergency care of Sickle Cell Disease patients

Color	Severity	Assistance needed	Complications / Diagnosis	Treatment
Red	High	Need immediate assistance in emergency service	Risk of death or irreversible injury: Acute Chest Syndrome, spleen sequestration, intense painful crisis, stroke, seizure	Intravenous medication and/or emergency transfusion + observation for at least 24 hours
Orange	Medium	Need for care in emergency service	Complaints and physical changes that need treatment in less than six hours, which can develop to health problems if untreated: Nonspecific infectious processes, priapism, moderate painful crises	Intravenous medications and/or non-emergency transfusion + need for observation for more than 12 hours
Yellow	Low	Need for care in emergency service not specializing in Hematology	Complaints and physical changes that need treatment in less than 12 hours that may evolve if not treated: Vomiting, pain, fever, respiratory infections	Venous medications
Green	Without	Care in outpatient service or health clinic (low complexity)	Complaints with little clinical relevance and without association with underlying disease: Colds, rubella, chickenpox, mild allergies, mild gastrointestinal disturbances, upper respiratory infection	Oral medication
Blue	Without	No need for emergency service	No clinical complications	No need for treatment

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 2 - Risk classification of complications of the selected sickle cell disease patients which resulted in visits to the Coordinating Blood Bank Emergency Department

Severity of complaint	2007		2008		Total	
	N	%	N	%	N	%
No complication	31	26.5%	26	32.1%	57	28.9%
Not severe	42	35.9%	27	33.3%	69	34.8%
Low severity	18	15.4%	10	12.4%	28	14.1%
Medium severity	10	8.5%	9	11.1%	19	9.6%
High severity	16	13.7%	9	11.1%	25	12.6%
Total	117	100%	81	100%	198	100%

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

Table 3 - Main complaints of sickle cell disease patients

Main Complaint	2007		2008		Total	
	N	%	N	%	N	%
Fever	37	31.6%	34	42.0%	71	35.8%
Pain	22	18.8%	8	9.9%	30	15.1%
Vomiting / Diarrhea	12	10.2%	8	9.9%	20	10.1%
Fatigue / pallor	10	8.5%	8	9.9%	18	9.1%
Outpatient care	11	9.4%	4	4.9%	15	7.6%
Respiratory symptoms	3	2.6%	6	7.4%	9	4.5%
Increased spleen	7	6.0%	1	1.2%	8	4.0%
Increased spleen	7	6.0%	1	1.2%	8	4.0%
Child crying excessively	3	2.6%	1	1.2%	4	2.1%
Avian influenza A (H1N1)	0	0.0%	1	1.2%	1	0.5%
Other complaints	9	7.7%	9	11.2%	18	9.1%
Total	117	100%	81	100%	198	100%

Table 4 - Most frequent clinical complaints reported by the selected sickle cell disease patients

Diagnosis	2007		2008		Total	
	N	%	N	%	N	%
No medical complication	30	25.6%	17	21.0%	47	23.7%
Upper respiratory tract infection	25	21.4%	15	18.5%	40	20.2%
Splenic sequestration	11	9.4%	7	8.6%	18	9.1%
Painful crisis	9	7.7%	5	6.2%	14	7.1%
Anemia	4	3.4%	5	6.2%	9	4.5%
Gastroenteritis	3	2.6%	4	4.9%	7	3.5%
Pneumonia	4	3.4%	2	2.5%	6	3.0%
Unspecified infection	4	3.4%	1	1.2%	5	2.5%
Cold	0	0.0%	5	6.2%	5	2.5%
Allergy	3	2.6%	2	2.5%	5	2.5%
Other diagnosis	24	20.5%	18	22.2%	45	21.4%
Total	117	100%	81	100%	198	100%

Table 5 - Outcomes of care provided by the Coordinating Blood Bank Emergency Department

Outcome	2007		2008		Total	
	N	%	N	%	N	%
Discharge	109	93.3%	74	91.4%	183	92.4%
Hospitalized to a ward	6	5.1%	7	8.6%	13	6.6%
Admitted to Intensive Care Unit	1	0.8%	0	0.0%	1	0.5%
Death	1	0.8%	0	0.0%	1	0.5%
Total	117	100.0%	81	100.0%	198	100%

Table 6 - Time of stay of sickle cell disease patients treated in the Emergency Room

Time of stay in the emergency room	2007		2008		Total	
	N	%	N	%	N	%
Up to 1 hour	57	48.7%	39	48.1%	96	48.5%
From 2 to 6 hours	23	19.7%	13	16.1%	36	18.3%
From 7 to 12 hours	5	4.4%	5	6.2%	10	5.0%
From 13 to 24 hours	11	9.4%	9	11.1%	20	10.0%
From 25 to 36 hours	1	0.8%	1	1.2%	2	1.0%
From 37 to 48 hours	14	11.9%	9	11.1%	23	11.6%
From 49 to 72 hours	3	2.7%	3	3.8%	6	3.1%
From 73 to 96 hours	0	0.0%	0	0.0%	0	0.0%
From 97 to 120 hours	1	0.8%	1	1.2%	2	1.0%
From 121 to 144 hours	1	0.8%	0	0.0%	1	0.5%
From 145 to 168 hours	0	0.0%	1	1.2%	1	0.5%
Not informed	1	0.8%	0	0.0%	1	0.5%
Total	117	100%	81	100%	198	100%

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 public funds to procedures that are really needed. Furthermore, early interventions may make a difference in the outcome.[18]

Using the adapted model to assess risk in sickle cell disease carriers in the Emergency Department,[15] we observed that 63.7% of treated cases had no signs that would justify emergency treatment. About 14% of cases were considered low risk, which means that these patients could have been treated in outpatient services or primary care facilities near their homes.

Only 44 (22.2%) visits were really considered emergency cases, but this did not indicate a necessity of treatment in a hospital specializing in hematology. These cases could be considered serious and should be treated in general emergency departments near to the patient's residence because of risk that might aggravate the status of the patient. This risk could even become lethal depending on the delay in clinical intervention in cases such as splenic sequestration, painful crisis and pneumonia.

Of 198 attendances in the Emergency Department, there was one death and 14 (7.0%) hospital admissions. These hospitalizations were related to only 8 patients with an average of 1.75 admissions for each patient. All were eventually discharged with an average of 8.92 days of hospitalization.

In a study conducted in the United States [19] published in 2010, the treatment of sickle cell anemia patients was monitored in several emergency healthcare facilities in the period from 1999 to 2007. A total of 197,333 visits were analyzed by gender, age, reason for treatment and outcome of care. The results showed, as expected, no significant difference between genders because this is not a autonomic (gender-linked) disease. However other variables were quite different from those found in this study, probably because all patients treated here are Brazilian National Health Service (SUS) patients and also because the average age of our patients was much lower, with a predominance of under 1-year-old children (78.2%) giving a mean age of approximately 3.5 years, while in the American study 72.7% patients were over 20 and only 12.2% were under 10 years old.

The most frequent reasons for care in the Rio de Janeiro State Coordinating Blood Bank Emergency Room were: fever, pain and gastrointestinal tract disorders (vomiting/diarrhea) making up 61% of the total. It was observed that 23.7% of all visits were free of clinical events and the most common diagnoses were upper respiratory tract infections (20.2%), splenic sequestration (9.1%) and painful crises (7.1%).

Regarding outcomes of visits to the emergency room, 92.4% of cases in this study were discharged directly from the emergency room, while in the American study this figure was 70.6%. The average length of stay of patients in the emergency room of the Blood Bank was 13.35 hours and in the American study this variable was not calculated.

The distance between the healthcare facility and residence is often quite far, which not only causes anxiety for relatives and the patient, but also delays the start of treatment of simple complaints that can then turn into more serious conditions, such as respiratory infections evolving to acute chest syndrome because of lack of adequate hydration and antibiotics. Moreover, this distance between residence and treatment may be a factor to abandon treatment because of economic, social and physical difficulties in the transportation of patients.

As primary healthcare is considered the working basis for all 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.[20]

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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