SICKLE CELL ANEMIA: CHARACTERIZATION OF THE PATIENTS ATTENDED IN A SPECIALIST OUTPATIENT CENTER*

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ABSTRACT: This qualitative research aimed to describe the clinical, social and demographic profile of patients with sickle cell anemia attended in a specialist outpatient center for Pediatric Hematology in Curitiba, Paraná. Between the 1st and 19th of October 2012, all the medical records of persons with hemoglobinopathies in the outpatient center were sought; 58 persons were identified who met the inclusion criteria. Data from case histories and clinical tests, from the first attendance up to the date of collection, were organized in categories. There was a predominance (76%) of children (1 to 12 years old), originating from 14 health regions in the state (70.5%), diagnosed in the first year of life (80%), monitored for three years or more (86.2%), with clinical signs of skin paleness (94.8%) and fever (93.1%), with pain crisis as the main complication (70.7%). The results may support planning and operations in the care of the patient with sickle cell anemia and her family members in this outpatient service and also in other similar services.
DESCRIPTORS: Chronic illness; Sickle cell anemia; Public health nursing.

ANEMIA FALCIFORME: CARACTERIZAÇÃO DOS PACIENTES ATENDIDOS EM UM AMBULATÓRIO DE REFERÊNCIA

RESUMO: Pesquisa de abordagem quantitativa que objetivou descrever perfil clínico, social e demográfico de pacientes com anemia falciforme atendidos em ambulatório de referência para Hematologia Pediátrica, em Curitiba/Paraná. Buscaram-se, de 1 a 19 de outubro de 2012, todos os prontuários de hemoglobinopatas do ambulatório e foram identificados 58 que atendiam aos critérios de inclusão. Dados de anamneses e exames clínicos, desde o primeiro atendimento até a data da coleta, foram organizados em categorias. Com predomínio (76%) de crianças (1 a 12 anos), procedentes de 14 regionais de saúde do estado (70,5%), diagnosticadas no primeiro ano de vida (80%), acompanhados há três anos ou mais (86,2%), com sinais clínicos de palidez cutânea (94,8%) e febre (93,1%), e como complicação principal a crise álgica (70,7%). Os resultados podem subsidiar o planejamento e ajustes no cuidado ao paciente com anemia falciforme e seus familiares neste serviço ambulatorial e também em outros semelhantes.
DESCRITORES: Doença crônica; Anemia falciforme; Enfermagem em saúde pública.

ANEMIA FALCIFORME: CARACTERIZACIÓN DE LOS PACIENTES ATENDIDOS EN UN AMBULATORIO DE REFERENCIA

RESUMEN: Investigación de abordaje cuantitativo cuya finalidad fue describir perfil clínico, social y demográfico de pacientes con anemia falciforme atendidos en ambulatorio de referencia para Hematología Pediátrica, en Curitiba/Paraná. Fueron investigados, de 1 a 19 de octubre de 2012, todos los prontuarios de hemoglobinopatas del ambulatorio e identificados 58 que atendían a los criterios de inclusión. Fueron organizados en categorías los datos de anamnesis y exámenes clínicos desde el primer atendimiento hasta la fecha en que fueron obtenidos. Con predominio (76%) de niños (1 a 12 años), procedentes de 14 regionales de salud del estado (70,5%), diagnosticados en el primer año de vida (80%), acompañados por tres años o más (86,2%), con señales clínicos de palidez cutánea (94,8%) y fiebre (93,1%), y como complicación principal la crisis de dolor intenso (70,7%). Los resultados pueden subsidiar el planeamiento y ajustes en el cuidado al paciente con anemia falciforme y sus familiares en este servicio ambulatorial, así como en otros semejantes.
DESCRIPTORES: Enfermedad crónica; Anemia falciforme; Enfermería en salud pública.

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INTRODUCTION

The hemoglobinopathies, in particular sickle cell disease (SCD), are among the genetic diseases with the greatest epidemiological importance in the world, and occur predominantly among those of African descent[1-3]. It is estimated that approximately 7% of the world population has some type of hemoglobinopathy, and that approximately 300,000 to 400,000 children with these disorders are born each year – among whom, 250,000 are SCD[4]. SCD is most frequent in populations on the African continent, in particular, in the equatorial and sub-Saharan regions, located to the north of the Kalahari desert. It stands out that in Angola (Africa), it can reach 37% of births[5].

There are population groups in the countries of the Americas with a large quantity of persons with sickle cell traits and high incidence of SCD[5]. In Brazil, there are more than 7 million persons carrying the gene for hemoglobin S (Hb S) in the heterozygous state (referred to as carriers of the sickle cell trait), with prevalence in the general population of 2 to 8%[3] and from 25,000 to 30,000 with SCD[6]. In Paraná, in the period 2002 – 2004, the prevalence of SCA was of 2.2:100,000 live births, with the largest number of cases in the 2nd Health Region – Metropolitan, and in the 15th Region – Maringá[7].

SCD is spread heterogeneously in the Brazilian population, with greatest prevalence in those states with the highest concentration of people of African descent, among the poorest social class, and with a mean life expectancy for adults below 50 years old. The rate of lethality among children below five years old, without the care necessary and stipulated, is of 80%[1], a situation explained by the incidence of serious acute events in childhood, but with evidence of concentration of deaths among those below two years old[3].

It is emphasized that the term SCD is generic. It refers to a group of genetic alterations whose characteristic is the predominance of Hemoglobin S (Hb S) and which includes sickle cell anemia (Hb SS), as well as associations of Hb S with the other variants of Hemoglobins and those interacting with thalassemias[8].

Clinically speaking, sickle cell anemia (SCA), as a chronic inflammatory disease, corresponds to the homozygatous state for Hb S, and is characterized by repeated episodes of vaso-occlusion, which are caused by the sickled red blood cells (sickle-shaped) and results in various clinical complications which affect all the organs and/or tissues[9], such as intense pain crises, fever, repeated infectious situations, splenic sequestration, cerebrovascular accidents (CVA), acute chest syndrome (ACS), and organ insufficiency, among others[2,10].

It is understood that the clinical course of SCA occurs with unique characteristics and, above all, the recurrence of clinical manifestations correlated with variables such as age and socioeconomic conditions is expected. Various countries, therefore, with high prevalence of SCD, have established comprehensive actions relating to the availability of the following components: emergency treatment, hospital for inpatient treatment, outpatient/community care, as well as the key strategy of reliable and comprehensive neonatal triage, with efficacious provision of ideal care for those with SCD[11-12].

It is emphasized that the aim of the neonatal triage program is to promote the diagnosis of congenital pathologies for children born alive, in the presymptomatic phase, and thus to provide appropriate treatment, reflected in the reduction of morbidity and mortality caused by the diseases for which people are triaged[7].

In this regard, in 2001 the Brazilian Ministry of Health institutionalized the National Neonatal Triage Program (PNTN), the "heel prick test", through Ministerial Ordinance N. 822. In Phase II of the Program, this included – mandatorily – triage for the hemoglobinopathies, that is, hemoglobin electrophoresis. Thus, all Brazilian newborns have come to have equal access to the test, regardless of their ethnic background, geographical origin, or socioeconomic-cultural status[2,6,13]. Prior to the PNTN, 20% of children with SCA did not reach five years of age, and the others showed marked reduction in school performance[7].

In Paraná, State Law 867/1987 made the "heel prick test" mandatory. This consists of triage for phenylketonuria in all newborns of the state. The services collaborating in this are the Pediatric Hematology Outpatient Center, of the Federal University of Paraná's Teaching Hospital, in Curitiba, and the entire Hemepar Network, which has 22 units distributed throughout the state of

Paraná, Brazil(7,14). Recently, in December 2013, the Study for Hemoglobinopathies in Pregnant Women – the "Mother's test" – began in the state of Paraná[14].

Thus, with the aim of supporting the care offered in the pediatric hematology service, it was sought to respond to the question: What are the social, demographic and clinical characteristics of the population attended in a specialist outpatient center for the treatment of sickle cell anemia? Therefore, this study aimed to identify the clinical, social and demographic profile of patients with sickle cell anemia, of the Pediatric Hematology Outpatient Center, a center of excellence in the state of Paraná, Brazil.

METHOD

This is a quantitative study with a descriptive nature. The study was undertaken based in records of attendance in the medical records of the clients registered and active in the Pediatric Hematology Outpatient Center, of the Teaching Hospital of the Federal University of Paraná (UFPR), in Curitiba, in the state of Paraná.

Data collection, undertaken between the 1st and 19th October 2012, was based on a search, and exploratory reading, of all the medical records (n=258) archived in the outpatient center, and were of patients with hemoglobinopathies. Half (n=129) were excluded, due to being carriers of the sickle cell trait (Hb AS); or carriers of trait C (HbAC); or presented other hemoglobinopathies (Hb SC, Hb SD, S/ thalassemia; hemoglobin C/ thalassemia; hemoglobin D/ thalassemia and thalassemia major).

Of the 129 medical records of patients with SCA, it was detected that 65 (50.4%) were not monitored in the outpatient center, and that six (4.6%) had died, these therefore being excluded. A total of 58 (45%) medical records remained, of people who were being clinically monitored – and this formed this study's sample.

Data were selected and extracted from medical histories and clinical tests, recorded in the 58 medical records, over the entire period of treatment of each patient, up until the date on which the data was collected. The following variables were selected: age (in complete years); sex; origin by Health Region of the State of Paraná; type of housing (masonry, wood); basic sanitation; parents' professional occupation, educational level and genotype; the age at which diagnosis was undertaken; length of time of monitoring in the outpatient center; clinical manifestations and complications; and causes of episodes of inpatient treatment associated with SCA.

The information obtained was organized in an electronic database, in Microsoft Excel, Windows 2007, and was subjected to descriptive analysis, measured by tables of absolute and relative frequency. This study is part of a line of research which was approved by the Ethics Committee, under Opinion N. 84,718, of 29th August 2012, of the Health Sciences Department of the UFPR.

RESULTS

In relation to the social and demographic profile of the carriers of SCA, the majority (76%) of the patients were in the age range between one and 12 years old (variation of one to 22 years old); 53.5% were female; 66.7% lived in houses built of masonry; 87.8% had basic sanitation; 80% had been diagnosed prior to completing one year of age, and 86.2% had been monitored periodically for three years or more in the outpatient center under study (Table 1).

Regarding the parents' social variables, such as educational level and professional occupation, the high unavailability of information recorded in the medical records did not evidence results which it was possible to analyze. In the few records available, the researchers noted the prevalence of junior high school not completed for both parents, and that the mothers were housewives (Table 2).

In relation to the parents' genotypes, apart from the records for 17 mothers and 24 fathers, whose absence was significant, it is ascertained that all the mothers and nearly all the fathers (97.06%) were carriers of the sickle cell trait, that is, they were heterozygotes for Hb S (Hb AS) (Table 2).

Approximately 71% of the patients were from other Health Regions of the state of Paraná, 38% from the North-Northeast regions; and 29.4% were from the 2nd Region - Metropolitan, the headquarters in Curitiba, where the outpatient center is located. The other states, with 5.9% [3] of the patients, were São Paulo and Santa Catarina (Table 3).

Among the clinical manifestations, skin paleness and fever were the most evident signs, close to 95% and 93% respectively. More than 60% of all the records presented situations of pain in upper limbs (MMSS), lower limbs (MMII) and in the abdomen, as well as jaundice and vomiting (Table 4). It is worth noting that the occurrence and reoccurrence of various complications was quite common in the same patient with SCA throughout the period in which he or she was monitored. The most common were pain crisis in nearly 71% of the individuals, followed by recurrent infections in more than 50%. Some of these had repercussions for the reasons for inpatient treatment ascertained over the clinical evolutions, predominantly the pain crises (74.1%) and pneumonia (46.6%).

Table 1 - Social and demographic profile of the patients with SCA attended in a specialist outpatient center. Curitiba, Paraná, 2012

Age range	**n**	**%**
1 to 4 years old	17	29,3
5 to 7 years old	13	22,4
8 to 12 years old	14	24,3
13 to 17 years old	11	18,9
18 to 22 years old	03	5,1
Sex		
Male	27	46,5
Female	31	53,5
Type of residence		
Masonry	28	66,7
Wood	14	33,3
Data unavailable	16	-
Basic sanitation		
Public network	36	87,8
Well/spring and cesspit	05	12,2
Data unavailable	17	-
Age at diagnosis		
< 1 year old	44	80
1 to 4 years old	07	12,7
5 to 7 years old	03	5,5
8 to 13 years old	01	1,8
Data unavailable	03	-
Length of monitoring in the Service		
<1 year	01	1,7
1 to 2 years	07	12,1
3 to 5 years	23	39,7
6 to 9 years	20	34,5
10 to 13 years	07	12

*For each variable, 58 medical records were consulted.
** Considered 100% for each variable.

Table 2 - Social and genotype profile of the parents of the patients with SCA attended in the specialist outpatient center. Curitiba, Paraná, 2012

	Mother		**Father**	
Parents' educational level	**n**	**%**	**n**	**%**
Junior high school incomplete	10	83,4	04	36,3
Junior high school	01	8,3	02	18,2
Senior high school incomplete	00	-	03	27,3
Senior high school	01	8,3	02	18,2
Data unavailable	46	-	47	-
Parents' professional occupation				
House wife/husband	21	84	-	-
Other professions***	03	12	07	33,4
Rural worker	**01**	**4**	**05**	**23,8**
Builder	-	-	05	23,8
Security staff	-	-	02	9,5
Mechanic	-	-	02	9,5
Data unavailable	33	-	37	-
Parents' genotypes				
AS (sickle cell trait)	41	100	33	97,1
SS (sickle cell disease)	0	-	01	2,9
Data unavailable	17	-	24	-

*For each variable, 58 medical records were consulted.
** Considered 100% for each variable.
***Other professions. Mother: attendant, student, maid. Father: builder's mate, industrial worker, assembly line worker, handy-man, butcher, footballer, military police officer.

Table 3 - Distribution of the patients with SCA by Health Region of origin. Curitiba, Paraná, 2012

Health Region	n	%
2nd Curitiba	15	29,5
7th Pato Branco	06	11,9
11th Campo Mourão	05	9,9
12th Umuarama	03	5,9
14th Paranavaí	03	5,9
3th Ponta Grossa	02	3,9
15th Maringá	02	3,9
16th Apucarana	02	3,9
18th Cornélio Procópio	02	3,9
19th Jacarezinho	02	3,9
20th Toledo	02	3,9
5th Guarapuava	01	1,9
9th Foz do Iguaçu	01	1,9
10th Cascavel	01	1,9
21st Telêmaco Borba	01	1,9
Other States	03	5,9
Data unavailable	07	-
Total	58	100

Table 5 - Distribution of the clinical complications and causes for inpatient treatment of the patients with SCA attended in the outpatient center. Curitiba, Paraná, 2012

Clinical complications	n*	%**
Pain crisis	41	70,7
Recurrent infections	31	53,4
Splenic sequestration	17	29,3
Anemia	14	24,1
Dactylitis	10	17,2
Cholelithiasis	9	15,5
Osteomyelitis	5	8,6
Ulcers in MMII	5	8,6
Cerebrovascular accident	4	6,9
Priapism	2	3,4
Total	138	-
Causes for inpatient treatment		
Pain crisis	43	74,1
Pneumonia	27	46,6
Splenic sequestration	11	19
Splenectomy	10	17,2
Infections of the upper airways	10	17,2
Cholecystectomy	5	8,6
Urinary tract infection	4	6,9
Cerebrovascular accident	4	6,9
Total	114	-

Table 4 - Distribution of the clinical manifestations found most frequently among the patients with SCA attended in the specialist outpatient center. Curitiba, Paraná, 2012

Clinical manifestations	n*	%**
Skin paleness	55	94,8
Fever		93,1
Cough	38	65,5
Pain MMSS	37	63,8
Abdominal pain	37	63,8
Pain MMII	36	62,1
Jaundice	35	60,3
Vomiting	35	60,3
Edema MMSSII	34	58,6
Clear nasal discharge	32	55,2
Chest pain	27	46,5
Intolerance to physical activity	25	43,1
Diarrhea	25	43,1
Lower back pain	24	41,4
Dyspnea	20	34,5
Lymphadenomegaly	20	34,5
Asthenia	16	27,6
Splenomegaly	11	19
Hepatomegaly	08	13,8
Convulsions	03	5,2
Total	572	-

*In relation to the number of times they appear in the medical records consulted (n=58), even if these re-occur in each consultation.
**Proportionality calculated based on the total of medical records, that is, on the total of patients studied (n=58).

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DISCUSSION

Knowing the developmental periods of the patients and their relationship with the evolution of the SCA allows the health professionals to establish more accurate care conduct, as due to the seriousness of the hemolytic process, early deficits occur in weight and height, as do delays in sexual maturation, in children and adolescents. There is also evidence of intellectual impairment and subtle neuropsychiatric deficiencies, simply resulting from factors associated with a chronic illness, such as frequent episodes of hospitalization, missing school, socioeconomic difficulties and subclinical cerebral lesions, caused by repeated episodes of vaso-occlusion[15-16].

The prevalence of children below 12 years old may reflect the neonatal diagnosis through the compulsory triage, associated with the rapid system of referring these children to a specialized service considered a center of excellence in the state. There is a scarcity of publications addressing the sex of patients with SCA, due perhaps to the fact of this being a genetic illness which is not linked to sex; the greater prevalence of females may reflect the Brazilian population, in which women predominate[17].

Factors which are acquired or environmental, as well as those which are hereditary, exercise an important influence on the physiological consequences in the course of the SCA. Generally speaking, the socioeconomic and educational level occupy a prominent position in determining variants such as access to medical attention and better housing and working conditions[18], with proportional quality of life. However, a study undertaken in Uberaba, Minas Gerais, with 47 carriers of SCA from the regional blood center and with individuals with SCA, evidenced a rise in the educational level, with senior high school complete in nearly 43% of the patients. However, the reality was not the same for the patients' socioeconomic condition, evidenced by the type of work (30% with no defined profession), residence in the outskirts of the city (74%) and low pay (from 50% of one minimum salary up to one minimum salary)[19].

In this study, more than half of the users of the outpatient center live in masonry houses, with basic sanitation from the public network in nearly 90% of the housing; hence a scenario was found which contributed to a better prognosis for the patients. This information is important, as water contaminated by nitrites and microorganisms originating from human and animal waste is a factor which accentuates morbidity and mortality in this clientele[16]; according to the literature, there is greater susceptibility to early destruction of the erythrocytes in environments with pollutant gases, and food and water contaminated by nitrites[16,18].

In another aspect, the diagnosis made prior to the first year of life being completed results from the Brazilian public health policy instituted in the ambit of the Unified Health System (SUS), since 2001, in which the "heel prick test" allows the diagnosis of hemoglobinopathies in newborns[11-13].

The Neonatal Triage for hemoglobinopathies has brought immeasurable benefits for the children and their families, as the altered result in the newborn functions as a sentry-event, triggering a cascade of tests in the other members of the family, with therapeutic recommendations and referrals where necessary. The PNTN, for the sickle cell diseases, regarding support and early monitoring for individuals without restriction by class or ethnicity recognisably respects equality in the SUS[8]. Nevertheless, the task of maintaining equality of access and high-quality attendance to all patients with SCA in all countries still requires efforts[11-12].

In this study, it was ascertained that nearly all the parents were asymptomatic heterozygotes (Hb AS), which is important genetic information in order to detect families which have a risk of producing children with SCD and to provide access to genetic counseling in the family planning[1,4].

The percentage of patients originating from other Health Regions, at approximately 71%, confirms that the outpatient center in healthcare for people with hemoglobinopathies is the state's center of excellence. However, it is argued that territorial distance can worsen appropriate adherence to the treatment, considering that outpatient accompaniment must take place periodically for the patient's entire life[16]. It is also highlighted that the clientele's origin in the North-Northeast regions (38%) of the State of Paraná may be explained historically due to the greater extent of African miscegenation existent in these regions[7,9].

The vast majority of the patients was accompanied periodically in the outpatient center for three years or more, which presupposes regular monitoring and preventive guidance, contributing to better quality of life and – as a consequence – a lower rate of mortality, principally from infectious problems in children younger than five years old, as evidenced in other studies[9,20].

The pain crisis, as the most dramatic clinical event of SCA, caused by microvascular occlusion with subsequent tissue damage, in response to the sickling of the red blood cells, represented the most frequent clinical complication in this study, and also the principle cause of the episodes of inpatient treatment. This complication begins unexpectedly and exercises a direct impact on the patient's quality of life, as it can behave chronically, with transitory and moderate episodes, lasting for between five and 10 minutes, through to generalized episodes, which last for days or weeks and require inpatient treatment[10, 12].

Fever was the second most evident clinical finding, and may be a manifestation derived from the pain crisis. It generally occurs as a result of tissue ischemia and liberation of endogenous pyrogens; secondary to the crises of hemolytic anemia or as a sign of infection. The recurrent infections, as the second most common clinical complication, are explained by splenic dysfunction, resulting from loss of the reticulo-endothelial function of the spleen, as a consequence of the recurrent splenic infarctions, which are promoted by this organ's microvascularization, precipitating the process of sickling, and with this, tissue hypoxia[10,20]. Some clinical findings present in the clientele assisted – vomiting, diarrhea, lymphadenomegaly, dyspnea, cough and clear nasal discharge - can reaffirm the recurrence of these infectious processes.

Splenic sequestration, characterized by the rapid and progressive drop in the levels of hemoglobin, represented the third most frequent clinical complication, responsible for nearly 19% of the causes of episodes of inpatient treatment. This event requires immediate intervention, as it can progress to hypovolemic shock. The splenectomy, which represented 17.24% of all causes of inpatient treatment, is the therapeutic treatment for this clinical complication, indicated after two crises of splenic sequestration or after the first serious episode[15,10,21].

Skin paleness affected nearly all patients with SCA, and is generally associated with jaundice, dyspnea, asthenia and intolerance for physical activity. It may be related to the lower survival of the red blood cells and, therefore, to hemolytic anemia. However, the manifestation of these symptoms becomes evident only in some cases, when the levels of hemoglobin suffer a marked reduction, as the majority of the patients manages to maintain chronic levels of hemoglobin between 6.7 and 7.5g/dl, with compensated anemia[10,16].

Cholelithiasis is the most common digestive complication, resulting in particular from chronic hemolysis, which promotes the development of gallstones. Its incidence increases with age, affecting more than 50% of the adult individuals with SCA, but 15.5% of the clientele in this study presented this type of organic dysfunction. The cholecystectomy aims to prevent possible complications such as perforation of the gallbladder, bile peritonitis, choledocholithiasis, pancreatitis and sepsis[10,15].

Although the rate of 6.9% of CVA is low, when compared to the other clinical complications associated with SCA, it is significant, as its recurrence after the first event is high, and in the absence of treatment, it reoccurs in two thirds of cases[10,15].

It stands out that this study may have been the impaired by the lack of records of the life context of the patients with SCA, with lack of information regarding parents' educational levels and professional occupations, and their living conditions.

FINAL CONSIDERATIONS

The results presented are useful for guiding the professionals in identifying the determinants of the clinical situation of the patients with SCA in any health service. In order to plan their actions, health professionals must investigate the clinical history reported by the patient and family, with reference to their lifestyle and the environment in which they live.

The absence of essential records must be analyzed in relation to the quality of the care. Thus, it is evidenced that it is imperative to record the specific characteristics of each patient and their families in the medical records, so as to benefit the planning of the health education program

for the same; as well as for the professionals' continuous education.

This approach to epidemiological knowledge requires the development of further research, such as, for example, the recognition and knowledge of the families' and professionals' expectations that programs promoting the health of patients with SCA in the service may be initiated.

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