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Sickle cell disease on children: a descriptive study

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ABSTRACT
Aim: We intend to describe the socioeconomic and demographical characteristics of children between the ages of 0 and 11 years old, diagnosed with sickle cell disease, and describe the attention given to these children in the city of São Francisco do Conde, Brazil. **Method:** This is a descriptive and quantitative study, based on a population of 15 children with sickle cell disease. The demographic, socioeconomic and clinical characteristics were collected through the use of a form containing structured questions, applied to the mothers/tutors of the children. The statistical analysis of the data collected involved the use of unvaried distribution of frequencies presented on the charts. **Results:** From the total of the population, 86.7% are afro-decedents; 33.3% lived with a net income of two minimum wages or less and; 73.3% has used the emergency system and was hospitalized. **Discussion:** The socioeconomic data of the identified population allow us to make inferences about the quality of the assistance given to the child living with the disease. **Conclusion:** The implication of the sickness could be minimized by healthcare actions that are consistent with the needs of these children.
Keywords: Child Health; Anemia, sickle cell; African Continental Ancestry Group

INTRODUCTION

Sickle cell disease is one of the most frequent genetic mutations in Brazil. This term defines a group of hemoglobinopathies that are characterized by the predominance of the hemoglobin (Hb) S in the erythrocytes. Among these is found sickle cell anemia (Hb SS), Hb SC, S-thalassemia, and other even rarer diseases, such as Hb SD and Hb SE[1,2].

The hemoglobin S, when without oxygen, suffers alterations in its physical-chemical proprieties which result in an alteration of the erythrocytes to a sickle format. This is one of the main pathophysiological mechanisms of the disease[1,3]. The vase-occlusive phenomenon and chronic hemolysis are the main determinants of the clinical manifestations[4].

Sickle cell disease is distributed heterogeneously throughout the population due to racial miscegenation, presenting a higher prevalence in areas in which the afro-descendent population is higher. Gene frequencies vary from 2% to 3% within the country as a whole, rising to between 6% to 10% among afro-decedents. Consequently, this sickness is commonly found in the North and Northeast regions of Brazil, where the arrival of Africans was more intense, and helped to build the local ethnic constitution[5].

According to estimates of the Brazilian National Program of Neonatal Screening, every year 3,500 babies are born with sickle cell disease[5,6]. Twenty per cent of them will not live to five years of age due to complications directly involved with the sickle cell disease[6]., Without a doubt, such a scenario allows the authorities to treat this pathology as a public health issue[7].

According to the data from the neonatal screening, the Brazilian state of Bahia presents the highest incidence of sickle cell disease, in the form of one case to every 650 births, and one bearer of the sickle cell trace to every 17 births[2].

Based on the data from the Brazilian Institute of Geography and Statistics (IBGE, in Portuguese), in 2012, in the city of São Francisco do Conde, which is located in the metropolitan region of the city of Salvador, the capital of the state of Bahia, is where the proportion of the population consisting of afro-decedents reaches 90.9%[8]. It is also estimated that this municipality presents a high incidence of sickle cell disease.

There is no cure for sickle cell disease. However, there are certain treatments that provide the patient with the right of a life of quality, and proportionate to their families' comfort and safety[9]. Prophylactic care represents the essence of the treatment until they reach five years of age, the period during which there are many cases of obit and serious complications[10]. Therefore, an early diagnose through the Guthrie test and proper treatment plays a fundamental role in the reduction of the morbidity and mortality rates of these children[5].

It is important to highlight that the ethnical group involved is mostly afro-decedent, at the bottom of the social pyramid, and presenting the worst epidemiological, educational and economic indicators[11]. In this context, the socioeconomic factors contribute significantly to the clinical variables and to the prognostic of people with sickle cell disease. These include such aspects as poor living conditions, insufficient quality and quantity of food, and a shortage of medical care, all factors that are mostly associated with low income and low education[12].

Considering the gravity and frequency of the sickle cell disease, and way it interferes in the quality of life, the specific objectives of this study were to a): identify the children between the ages of 0 and 11 years with sickle cell disease; b) describe the socioeconomic and demographic characteristics of the children identified, and that of their mothers and/or carers and; c) describe the healthcare service provided to them in the municipality of São Francisco do Conde, Brazil.

METHOD

This is a descriptive study which has adopted a quantitative approach, performed in the municipality of São Francisco do Conde, Brazil.

The population of this study consisted of 15 children diagnosed with sickle cell disease. They were between the ages of 0 and 11 years, residents in the city of São Francisco do Conde, whose mothers/carers agreed to participate in this research as informants of the study. This number, in spite of the fact it looks small, is extremely significant when considering the estimated statistics for this disease in Brazil, which shows one infected

person for every 1,000 (1:1,000) born alive. In the state of Bahia, this rate is 1:650. Considering the population of de São Francisco do Conde, of 33,183 inhabitants, and with a child population of approximate 5,551 individuals between the ages of 0 and 9 years, according to IBGE's Demographic Census of 2010, the proportion of 1:650 in this population corresponds to approximately eight infected children.

This study initially involved children between the ages of 0 and 5 years, but due to the small number of identified infected children, the age group was modified, and the age limit was set at 11 years of age. However, this age range is in accordance with the Brazilian Statute of Children and Adolescents, bill #8,069, passed on July 13th 1990, which considers as a child, a person who is less than 13 years of age.

The criteria for inclusion were: be between the ages of 0 and 11; reside in the city of São Francisco do Conde; be diagnosed with sickle cell disease and; the mother/carer needs to consent to participate in this study.

To identify these children with sickle cell disease, we undertook the following steps: we obtained a list of people diagnosed with sickle cell disease from the Association of Parents and Friends of the Disabled (APAE, in Portuguese), São Francisco do Conde branch; from this list, we selected a sample based on the targeted age group; we obtained contact information from the Family Health Clinics (USF, in Portuguese) so that we could communicate with the parents of the children identified with sickle cell disease (data collected from the historical records of the USFs and complemented with data gathered from community health agents).

17 children were identified with sickle cell disease. However only 15 were found from the contact information provided. Two addresses could not be found.

The data collection was performed in the residence of the child using a form consisting of structured questions. This was applied to the parents or carers of the children, but preferably with the mothers. This step occurred between February and March 2011. The variables included the demographic, clinical and socioeconomic characteristics of the children (gender, age, ethnicity, type of sickle cell disease, recognition of the breadwinner, education of the breadwinner, family income, economic class according to the Brazil Economic Classification Criteria proposed by the Brazilian Research Companies

Association[13], number of residents, number of rooms, type of residence and if there is piped water and a septic tank); demographic variables regarding the caretakers (gender, family bond with the child, age) and variables regarding the identification of the caring given to the child with sickle cell disease (accompanying services, frequency of these services, professionals in the Healthcare Service who are responsible for the child, vaccination booklet, medication used, regular palpation of the spleen, the typical behavior in the case of pain reported by the child, usage of emergency services, hospitalization cases and daily ingestion of water by liter, by the child).

After the data was collected, the information was inserted and checked in a data bank elaborated into the Microsoft Access platform. It was later exported to statistical software entitled STATA, version 8. The statistical analysis of the data involved the use of distribution by unvaried frequencies aimed at describing the socioeconomic and demographic characteristics.

This research is part of the project entitled "Factors of Vulnerability of Children and Adolescents Health in São Francisco do Conde", funded by the Foundation of Research Support from the State of Bahia (FAPESB, in Portuguese), approved by the Ethics Committee of the Nursing School of Bahia Federal University (EEUFBA, in Portuguese), under protocol 04.2010. During this study, we followed all proceedings regarding research involving human beings. Therefore, a Free and Clear Consent Term was signed by the mothers/carers of sickle cell disease children, the participants of this study.

RESULTS

15 children diagnosed with sickle cell disease were identified in São Francisco do Conde, in the period between February and March 2011. In Table 1, which presents the demographic and clinical characteristics of the children under consideration, we observe that 60% of the children are male and present sickle cell disease type SC. All the others present type SS, except for carer who could not inform us of her child's sickle cell disease type (6.7%).

Regarding the age, it was observed that 53.3% were in the six to eleven years' age group, followed by the age group composed of children between two and five years of

age (33.3%). Regarding the variable *ethnicity*, 86.7% of the children are black, and among those 60% are declared by their mothers or carers as such, and 26.7% are brown.

Table 1- General characterization of the children with sickle cell disease in São Francisco do Conde, February-March 2011 (n=15).

Characteristic	N (%)
Type of Sickle Cell Disease	
SC	9 (60.0)
SS	5 (33.3)
Were not able to inform	1 (6.7)
Gender	
Female	6 (40.0)
Male	9 (60.0)
Age Group	
< 2 years old	2 (13.3)
2 to 5 years old	5 (33.3)
6 years old or older	8 (53.3)
Color/Ethnicity	
White	2 (13.3)
Black	9 (60.0)
Brown	4 (26.7)

Source: Project *Factors of vulnerability in children and adolescents health in São Francisco do Conde* (FAPESB), July 2011.

Regarding the socioeconomic characterization of the children under consideration, it can be seen from Table 2 that the majority belong to Class C (46.6%), followed by Class B (26.7%). In the field of housing, 93.3% were homeowners who had access to piped water, and among those, 60% had a septic tank.

From the total of the studied population, 73.3% reside in a house with three or four individuals, while 26.7% lived with five or more individuals. In terms of the number of rooms per house, the following was observed as the most common setting: two bedrooms, a living room, a kitchen and a bathroom (40%).

Table 2 – Socioeconomic characterization of the fifteen children studied, São Francisco do Conde, February-March 2011 (n=15).

Characteristic	N (%)
Brazilian Criteria of Economic Classification	
B	4 (26.7)
C	7 (46.7)
D	2 (13.3)
E	2 (13.3)
Number of inhabitant in the residence	
3 to 4	11 (73.3)
5 or more	4 (26.7)
Amount of rooms	
1 bedroom, 1 living room, 1 kitchen, 1 bathroom	1 (6.7)
2 bedrooms, 1 living room, 1 kitchen, 1 bathroom	6 (40.0)
3 bedrooms, 1 living room, 1 kitchen, 1 bathroom	1 (6.7)
1 interspace	2 (13.3)
Others	5 (33.3)
Type of residence	
Owned	14 (93.3)
Others	1 (6.7)
Piped Water	
Yes	14 (93.3)
No	1 (6.7)
Sceptic Tank	
Yes	9 (60.0)
No	6 (40.0)
Breadfeeder	
The woman alone	9 (60.0)
The male partner	6 (40.0)
Education level of the breadfeeder	
Illiterate/Elementary incomplete	2 (13.3)
Elementary complete/Middle incomplete	7 (46.1)
Middle complete/ high incomplete	3 (20.0)
High complete/ College incomplete	2 (13.3)
College complete	1 (6.7)
Family income (in minimum wage - MW)	
Less than 1 MW	1 (6.7)
1 MW	2 (13.3)
More than 1 MW and less than 2 MW	5 (33.3)
More than 2 MW and less than 3 MW	4 (26.7)
More than 3 MW and less than 4 MW	3 (20.0)

Source: Project *Factors of vulnerability in children and adolescents health in São Francisco do Conde* (FAPESB), July 2011.

In Table 3 there is a description of the demographic characterization of the mothers/legal tutors of the children. From the 15 people interviewed, 14 were the mothers and one was the grandmother, and the majority (53.3%) were in the age group 30 to 35 years.

Table 3 – Demographic characterization of the mothers/legal tutors of the children, São Francisco do Conde, February-March 2011 (n=15).

Characteristic	N (%)
Gender	
Female	15 (100.0)
Relationship with the child	
Mother	14 (93.3)
Grandmother	1 (6.7)
Age group	
< 30 years old	5 (33.3)
30 to 35 years old	8 (53.3)
36 to 40 years old	1 (6.7)
41 years old or older	1 (6.7)

Source: Project *Factors of vulnerability in children and adolescents health in São Francisco do Conde* (FAPESB), July 2011.

Table 4 describes the caring provided to children infected with sickle cell disease. It is observed that all children were supported by a healthcare service, with the APAE Salvador branch (26.6%) and the Hematology and Hemotheraphy Foundation of Bahia (HEMOBA) (26.6%) being the most commonly mentioned. The children attended every six months (33.3%), followed by reports every four months (20.0%).

The only professional mentioned by all responders, with responsibility for the treatment of the child with sickle cell disease, was the doctor (100%). The treatment provided, most of the time, happens in the specialized areas of pediatrics (86.7%) and hematology (46.6%). Besides the doctors, other professionals were also mentioned as part of the multi-professional health team that aids the child, These were dentists (40%), social workers (26.7%), nutritionists (20%) and nurses (20.1%).

Regarding the vaccination process, 86.7% of the children presented an incomplete vaccination booklet. From the observed vaccines, 50% of the missing ones are the

Pneumococcal conjugate 7-valent. Other mentioned vaccines were Anti-Influenza and Anti-Hepatitis A, and others from the regular national vaccination schedule. It was found that, from the medication used, all children are prescribed with folic acid, and 80% use a frequent antibiotic, 75% use oral Penicillin V and 25% benzathine Penicillin.

The majority of the mothers/carer (73.3%) do not know how to perform the palpation of the child's spleen. In the event of pain, 93.3% of the mothers use analgesics and, from those, 13.3% use, besides analgesics, they looked for health services (13.3%).

It was noted that the forwarding of the children to health services identified that 73.3% of the mothers/tutors used the emergency services due to the incidence of intense pain (73.5%), followed by fever (53.5%). The higher percentage of children (73.3%) was already hospitalized, with 53.3% of them more than five times.

All mothers/carer mentioned that their children have a diversified diet and that the majority of them drink two or more liters of water per day (66.7%).

Table 4 – Caring provided to the child with sickle cell disease, São Francisco do Conde, February-March 2011 (n=15).

Caretaking	N (%)
Services used to assist the children with Sickle Cell Disease	
APAE Salvador branch	4 (26.7)
HEMOBA	4 (26.7)
APAE Salvador branch / PSF unit	1 (6.7)
APAE Salvador branch / Others	1 (6.7)
APAE Salvador branch / PSF unit / Others	1 (6.7)
HEMOBA/ PSF unit / Others	1 (6.7)
Others (Clinic - SFC, Hospital- SFC, Peterson - Salvador)	3 (20.0)
Frequency of usage of health services	
Monthly	1 (6.7)
Every 2 months	1 (6.7)
Every 3 months	3 (20.0)
Every 4 months	3 (20.0)
Every 5 months	2 (13.3)
Every 6 months	5 (33.3)
Professional that takes care of the child	
Pediatrician	2 (13.3)
Nurse / Pediatrician	1 (6.7)
Pediatrician / Dentist	1 (6.7)

Pediatrician / Others	1 (6.7)
Social worker / Others	1 (6.7)
Nurse / Pediatrician / Others	1 (6.7)
Pediatrician / Social worker / Dentist	2 (13.3)
Pediatrician / Social worker / Others	1 (6.7)
Pediatrician / Nutricionist / Dentist	1 (6.7)
Pediatrician / Nutricionist / Others	1 (6.7)
Pediatrician / Dentist / Others	1 (6.7)
Nurse / Pediatrician / Nutricionist / Dentist	1 (6.7)
Others (Hematologist, General Practicioner, Phychologist, Ophtalmic)	1 (6.7)
Vaccination booklet	
Complete	2 (13.3)
Incomplete	13 (86.7)
Folic Acid	
Uses	15 (100.0)
Routine use of antibiotics	
Uses	8 (53.3)
Does not use	7 (43.7)
Regular palpation of the child's spleen	
Knows how to do the procedure	4 (26.7)
Does not know how to do the procedure	11 (73.3)
Normal procedures in reported pain	
Uses analgesic	11 (73.3)
Does not do anything	1 (6.7)
Uses analgesic / Intensifies hydration	1 (6.7)
Uses analgesic / Searches health service	2 (13.3)
Use emergency services	
Yes	11 (73.3)
No	4 (26.7)
Hospitalization	
Yes	11 (73.3)
No	4 (26.7)
Number of hospitalizations	
Up to 5	7 (46.7)
More than 5	8 (53.3)
Daily ingestion of water, in liters, by the child	
Less than 2 liters	4 (26.7)
2 or more liters	10 (66.7)
Not able to respond	1 (6.7)

Source: Project *Factors of vulnerability in children and adolescents health in São Francisco do Conde* (FAPESB), July 2011.

With respect to the forms of prevention of complications, 86.7% of the informants mentioned the use of immuneprophylaxis, 66.7% use antibioticprophylaxis, 73.3% use

oral hydration, 66.6% protect the children with appropriate clothing to protect them from ordinary climate alterations and in 59.9% of the children are supported by a multi-professional health team.

DISCUSSION

The identification of the demographic, socioeconomic and clinical characteristics of the children with sickle cell disease is important to guide the caring procedures, and consequently promote the improvement in the quality of life of individuals with this pathology.

It is known that the disease under consideration does not discriminate, but a critical profile in terms of health and living conditions can be a determinant when it comes to acquiring, curing or rehabilitating the patient in the face of a worsening in general health conditions[14].

From the characterization of the children with sickle cell disease, it was seen that the majority of them were male and presented at the age of six years or older. The sickle cell disease diagnosed in this study was mostly of the SC type. This fact does not match with the studies of hemoglobinopathy prevalence undertaken in different regions of Brazil. As was described by Amorim et al. (2010), the two most frequent abnormal hemoglobins in the Brazilian population are hemoglobins S and C, predominantly the combination SS[15]. While correlating the presence of abnormal hemoglobins with the child's ethnicity, this study confirms the higher prevalence of this disease in the Afro-Brazilian population.

Regarding the characterization of mothers/carers, it was identified that they were all female, mainly between the ages of 30 and 35 years, and declared themselves as the main person responsible for financially supporting the family. The majority had attended Elementary and Middle schools, and were in receipt of a family income consisting of one and two minimum wages.

In a study performed in 2009, the patients with sickle cell disease are mostly among the Afro-Brazilian population. These are mainly poor and have difficulty in taking the sick child to the hospital. In these families, some members are illiterate, which interferes with

an understanding of the steps recommended by the health professionals, either in terms of healthcare procedures, or the laws that guarantee their rights[16].

Observing the housing conditions, it was seen that the majority of houses consisted of three to four residents living in five rooms. According to the Brazilian Criteria for Economic Classification, the majority of the subjects involved in this study belong to Class C. In terms of sanitation, only one house did not have piped water, and six did not have a septic tank. It is worth mentioning that the municipality under consideration does not have a sewage system. Therefore, the deficiencies in terms of housing and sewage conditions are elements usually associated with low income communities. This can interfere seriously with the physiopathology of the sickle cell disease, contributing to a situation of higher risk in terms of complications, especially infections. Furthermore, the housing condition of these children in São Francisco do Conde is a factor that contributes to their vulnerability.

With regard to the care treatment provided to the children, it was seen that the majority of the services used by them were at the APAE Salvador branch and HEMOBA, where they attended every six months. Other services mentioned during the interviews were the emergency and hospitalization services. This indicates that the clinical assistance provided was not sufficient to prevent the development of the disease.

The assistance to the individuals with sickle cell disease is generally seen as the responsibility of hematological centers. However, this assistance must be performed by the health network at all levels[17]. As such, it is necessary to prepare these services, as well as its professionals, in order to provide qualified and complete assistance, as is proposed by the Brazilian National Policy for Full Attention to People with Sickle Cell Disease[5].

All the children in this study used folic acid. We highlight the importance of this substance in order to avoid its deficiency in the organism, and consequently, the development of megaloblastic anemia[18]. With antibiotics and vaccination, we observed that the majority of the children under consideration constantly use the first, but are not vaccinated, as they should be. This is a good result, such as it is the importance of prophylaxis using Penicillin Benzathine or oral V, from four months of age to five years,

despite the fact that, in association, the importance of the special vaccination and the Brazilian basic vaccination calendar helps, significantly, to ensure a drastic reduction of incidence and mortality due to infections caused by encapsulated germs[19].

Regarding the palpation of the spleen, this study identified that the majority of the mothers/carer do not know how to palpate the spleen as a method of prevention of a splenic sequestration crisis. This fact contributes to the interference in the sudden rise of spleen volume and the search for medial assistance. This method involves a person using a wooden tongue depressor, with one of the extremities facing the umbilical scar and touching the edge on the palpable spleen. A movement should then be done, guiding the depressor against the left rib cage. Such a strategy does not require the use of a metric tape that can limit the use of such an approach on the part of some carers due to their illiteracy[2].

In terms of forms of prevention, the majority of mothers had an understanding of the importance of measures such as immunoprophylaxy, antibioticprophylaxy, hydration, use of proper clothing to cope with normal climate changes, followed by a multi-professional health team, and a diversified diet.

The parents' or tutors' education about the disease is extremely important. The relatives must be guided by the necessity to ensure the diseased child's hydration and proper nutrition, the importance of preventing infection aided by vaccines, the use of prophylactic penicillin and a recognition of the inter-occurrence of diseases[20].

CONCLUSION

In this study, we were able to identify that the number of children infected with sickle cell disease in São Francisco do Conde was above the expected, considering that the study examined an importante institution to identify the children who were diagnosed with sickle cell disease. Therefore, this data could also be an underestimate of the true situation due to the low coverage of neonatal screening within the municipality, which limits the identification of children with the pathology.

In addition, the data presented shows that, although São Francisco do Conde has an elevated *per capita* income, the population identified here presents a low socioeconomic status which interferes directly with the caring assistance given to children with sickle cell disease and, consequently, with their clinical condition.

It is also seen that the attendance of the children to health service facilities, and the caregiving by the mothers/tutors do not meet their needs. Therefore, the assistance of multi-professional health teams and the implementation of the Brazilian National Policy of Attention to People with Sickle Cell Disease in the municipality is a necessity.

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