

THE IMPACT OF SICKLE CELL DISEASE ON THE DAILY ROUTINE OF ADOLESCENTS*

Márcia Helena de Souza Freire¹, Rafaela Aparecida Pereira², Evelyn Juliana Ramos³, Vanilde de Fátima Andrade Matos⁴, Michelle Thais Migoto⁵

¹Nurse. PhD in Public Health. Professor at the Department of Nursing of Universidade Federal do Paraná. Curitiba, PR, Brazil.

²Nurse. Family Medicine Residency Program. Secretaria Municipal de Saúde de Curitiba. Curitiba, PR, Brazil.

³Nurse. *Fundação Estatal da Atenção Especializada à Saúde*. Curitiba, PR, Brazil.

⁴Nurse. Masters student in Psychology. *Ambulatório de Hematologia Pediátrica do Hospital de Clínicas* at UFPR. Curitiba, PR, Brazil.

⁵Nurse. Masters student in Nursing. Universidade Federal do Paraná. Curitiba, PR, Brazil.

ABSTRACT: The present study aimed to identify the impact of sickle cell disease on the daily lives of adolescents. Exploratory, descriptive, qualitative and quantitative research with 12 adolescents enrolled in an outpatient unit of the city of Curitiba, state of Paraná. Data collection was performed between January 15 and April 30, 2014. The theory of Social Representations was the reference of the research, and the technique used was Collective Subject Discourse methodology. Half of the adolescents were of African descent, and almost all of them were diagnosed at the National Newborn Screening Program – Guthrie test. The youngsters had some knowledge of the disease; most of them reported not feeling different from other adolescents not affected by the disease; there were frequent reports of pain, fatigue, medication use and limitations of social life; the most common reason for hospitalization was pain episodes. Educational actions in health services focused on the characteristics of the patients and their families, will minimize the impacts of the disease and promote the quality of life of adolescents with sickle cell disease.

DESCRIPTORS: Sickle cell disease; chronic disease; Adolescent; Nursing in public health; Qualitative research.

O IMPACTO DA ANEMIA FALCIFORME NA VIDA DE ADOLESCENTE

RESUMO: Objetivou-se identificar o impacto da anemia falciforme no cotidiano de adolescentes. Pesquisa qualitativa, descritiva e exploratória com 12 adolescentes inscritos num ambulatório da cidade de Curitiba, estado do Paraná. A coleta dos dados ocorreu entre 15 de janeiro a 30 de abril de 2014. Teve como referencial teórico as Representações Sociais e metodológico o Discurso do Sujeito Coletivo. Dos resultados encontrados, metade dos adolescentes era afrodescendente, quase todos foram diagnosticados na Triagem Neonatal no Teste do Pezinho. Os jovens demonstraram algum conhecimento da doença; a maioria referiu não se sentir diferente perante outros adolescentes sem a doença; foram frequentes os relatos de dor, cansaço, uso de medicamentos e limitações ao convívio social; o motivo de internação mais frequente foi por crise alérgica. Ações educativas em serviços de saúde, com foco nas singularidades de paciente e família, minimizarão os impactos da doença e promoverão qualidade de vida dos jovens com anemia falciforme.

DESCRIPTORES: Anemia falciforme; Doença crônica; Adolescente; Enfermagem em saúde pública; Pesquisa qualitativa.

EL IMPACTO DE LA ANEMIA FALCIFORME EN LA VIDA DE ADOLESCENTE

RESUMEN: Estudio cuya finalidad fue identificar el impacto de la anemia falciforme en el cotidiano de adolescentes. Investigación cualitativa cuantitativa, descriptiva y exploratoria con 12 adolescentes inscritos en ambulatorio de la ciudad de Curitiba, Paraná. Los datos fueron obtenidos entre 15 de enero y 30 de abril de 2014. El referencial teórico fue de las Representaciones Sociales y, el metodológico, del Discurso del Sujeto Colectivo. Acerca de los resultados, mitad de los adolescentes era afrodescendiente, casi todos fueron diagnosticados en la Selección Neonatal en el Test de Guthrie. Los jóvenes demostraron algún conocimiento de la enfermedad; la mayoría declara no sentirse diferente delante de otros adolescentes sin la enfermedad; fueron frecuentes los relatos de dolor, cansancio, uso de medicamentos y limitaciones del convivir social; el motivo de internación más frecuente fue por crisis de dolor. Acciones educativas en servicios de salud, con foco en las singularidades de paciente y familia podrán minimizar los impactos de la enfermedad y promover cualidad de vida de los jóvenes con anemia falciforme.

DESCRIPTORES: Anemia Falciforme; Enfermedad Crónica; Adolescente; Enfermería en Salud Pública; Investigación Cualitativa.

*Article extracted from the end-of course paper titled "*Anemia falciforme à luz da enfermagem: caracterização dos portadores, impacto pessoal e familiar, promoção em saúde*". Universidade Federal do Paraná, 2014.

Corresponding author:

Márcia Helena de Souza Freire
Universidade Federal do Paraná
R. José de Mello Braga Júnior, 143, 81540-280 - Curitiba, PR, Brasil
E-mail: marcia.freire@ufpr.br

Received: 07/07/2015

Finalized: 11/08/2015

INTRODUCTION

Sickle cell disease (SCD) is a chronic, autosomal recessive genetic disorder that affects hemoglobin S (HbS). It originated in different regions, particularly African communities and the Mediterranean Region, and is more common among individuals of African descent.. Therefore, ethnicity is relevant for this pathology⁽¹⁾, recognized as a priority by the World Health Organization (WHO), since it is considered a public health issue because of morbimortality in children under 5 years.

An international study that used the World Bank's Global Bilateral Migration Database showed an increase in the number of individuals with HbS as a result of the phenomenon of globalization and the consequent increase in immigration and miscegenation in countries of the European, American and Asian continents. The same phenomenon was observed in African countries⁽²⁾.

The estimated trend in the future behavior of sickle cell diseases is a rise in the overall number of newborns from 305,800, in 2010, to 404,200, in 2050. This increase in the number of births has been significant in Nigeria and in the Democratic Republic of Congo, and the opposite has been observed in India. In view of this scenario, the implementation of different levels of health intervention are recommended to save the lives of children with sickle cell disease (SCD) under five years of age⁽¹⁾, especially policies on genetic counseling and screening of populations⁽²⁾.

The individuals with SCD may have varied symptoms, with different manifestations. The most common manifestations such as pain, paleness and nausea, in addition to affecting many organs also cause emotional and social impacts that impair the quality of life of the patients⁽³⁾ and their families, particularly in the life cycle of childhood and adolescence.

In developed countries, the main causes of complications of children with SCD between 1 and 3 years of age are infections, acute splenic sequestration crisis and acute chest syndrome. Thus, we stress that early diagnosis and comprehensive treatment are actions that reduce mortality and increase survival in chronic patients⁽⁴⁾, though with a reduced life expectancy forecast, less than 45 years, and complications in most patients⁽⁵⁾.

In order to reduce the impact of SCD, priority

should be given to governmental policies to control the disease and to promote health education, especially in primary health care and in specialized centers.

Therefore, being aware of the difficulties and challenges faced by individuals with SCD may improve the training of health professionals and the handling of the adversities caused by the disease. The abovementioned provides justification for this research that attempted to identify the impact of SCD on the daily routine of adolescents.

METHODS

Descriptive qualitative and quantitative research conducted in an Outpatient Pediatric Hematology Clinic (*Ambulatório de Hematologia Pediátrica*), in the city of Curitiba, state of Paraná. It is a referral center in the state for monitoring and treating hemoglobinopathies. The collection of empirical data occurred from January 15 to April 30, 2014. The interviews used a semi-structured instrument and quantitative data for the characterization of the adolescents. The variables used in this characterization were age, ethnicity, gender, education, person who accompanied the adolescent, number of hospitalizations and reasons. The interviews included the following questions: What do you understand by sickle cell disease? Tell us about when and how your diagnosis was made. How do you feel about your colleagues? Does sickle cell disease interfere in your activities? Do you have any suggestions for this outpatient clinic?

Inclusion criteria were, as follows: 12-25 years of age and be in the waiting room on the days of the appointments established for data collection. The medical records of the participants were examined and the individuals who met the inclusion criteria were selected. The informed consent documents were signed and participants were free to decide to withdraw from the study at any moment. Twelve adolescents were interviewed.

The method of data analysis and presentation concerned the technique of Collective Subject Discourse (CSD)⁽⁶⁾, where the individual voice (statement) of the respondent expresses a collective voice. The CSD is based on the theory of Social Representation (SR), as it provides appropriate knowledge of the social reality shared favoring intervention⁽⁷⁾.

Using the software Qualiquantisoft®, we

analyzed the empirical substrate collected from the responses of the participants. For every response, key expressions that translated the essence of each segment of the response were elected. The core idea (CI) conveyed by each key expression was identified for the synthesis and grouping of these expressions. The election of the categories of analysis was done in an orderly manner, considering all the similar discursive segments that pointed to similar information or meanings. At the end of the methodological process, the synthetic discourses related to each category of analysis were obtained. Therefore, each CSD was defined based on the discursive fragments from all the responses to each question, the number of times the discourse appeared in the responses is quantified with the use of the software and expressed as the representativeness (sharing power) of the idea, in percentage⁽⁷⁾.

The study's accuracy was observed through the use of Consolidated Criteria for Reporting Qualitative Research (COREQ), a tool that supports the explicit and comprehensive report of qualitative studies. It is a 32-item checklist that systematically presents important aspects of the research team, methods and context of the study, findings, analysis and interpretations⁽⁸⁾.

The umbrella research that contemplates this study began after the approval of the Project by the Research Ethics Committees of the Health Sciences Sector and of the hospital to which the outpatient clinic is attached, under No 84718, of August 29, 2012.

RESULTS

Twelve adolescents aged 12-17 years old were interviewed, and eight of them were 12 years old; there was a predominance of female subjects (10); black ethnicity (6); elementary school students (10); five individuals from the 2nd Regional de Saúde - RS (regional health office) of the State of Paraná, which is headquartered in Curitiba; the participants were accompanied by their mothers (11); and were hospitalized due to pain episodes (10), as shown in Table 1.

The synthetic CSD were organized in categories with the aid of Qualiquantisoft®, according to the similarities of the core ideas (CI) quantified in the responses, shown with simple and relative frequencies, the later indicating the power of expression. Of the categories (Chart 1).

Knowledge about sickle cell disease

The participants reported having some

Tabela 1 - Caracterização sociodemográfica dos adolescentes com anemia falciforme, Ambulatório de Hematologia Pediátrica. Curitiba, PR, Brasil, 2014

Variables	N	%
Gender		
Female	10	83.3
Male	2	16.7
Age		
12 years old	8	66.7
13 years old	4	33.3
Ethnicity		
Black	6	50
Brown	3	25
White	3	25
Education		
Elementary school	10	83.3
High school	2	16.7
Origin		
2ª RS	5	41.7
Other RS	6	50
Another state	1	8.3
Escorts		
Mother	11	60.1
Other relatives	7	39.9
Hospitalizations		
Pain episodes	10	62.5
Other causes	4	24.9
Not hospitalized	2	12.6

knowledge about sickle cell disease. The responses were divided into two categories: lack of knowledge and knowledge. Ambos com 50% de força de compartilhamento.

Lack of knowledge: *"No... I don't know. At the other clinic there were lectures. The place was divided: there was a space for the children and one for the parents, and I stayed in the space for children. I've never got a clear explanation about it"* (DSC-A).

Knowledge: *"I know it affects my blood flow, is an incurable disease, my mother told me about it... I know it deforms blood cells.. the doctors are always talking about it... deformed cells throughout the body. I know there is a trait inherited. I know that people with SCD have more white than red blood cells. I learned it here and at school too"* (DSC-B).

Diagnosis of the disease

The respondents explained how their diagnoses were made. In 66.7% of the expressions diagnosis occurred during the Newborn Screening Program,

Chart 1 – Distribution of the questions and categories according to their frequency (n) and expression. Power (%) in the individual discourses, Ambulatório de Hematologia Pediátrica. Curitiba, PR, Brazil, 2014

Questions and Categories	N	%
Question 1: What do you understand by SCD??		
A. Lack of knowledge	6	50
B. Knowledge	6	50
Question 2: Tell me a little about when and how your diagnosis was made.		
A. At the National Newborn Screening Program	8	66.7
B. To investigate morbidities/hospitalizations	4	33.3
Question 3: How do you assess yourself as compared to your peers?		
A. Feels different from them	2	16.7
B. Does not feel different	10	83.3
Question 4: How does SCD interferes with your daily activities?		
A. Physically	10	62.5
B. Socially	3	18.7
C. Does not interfere	3	18.7
Question 5: What care/therapies are you receiving in this outpatient clinic?		
A. Medical appointments	11	47.8
B. Medication	12	52.2
Question 6: Do you have any suggestions for improvement of the services of this outpatient clinic?		
A. Expresses satisfaction	11	91.7
B. Dissatisfied with the disease	1	8.3

and in others it was made during the investigation of morbidities.

During Newborn Screening Program: *"I think the diagnosis was made soon after I was born. My mother learned of the disease during Guthrie Test... when I was four months old"* (DSC-A).

Through investigation of morbidities/hospitalizations: *"My mother told me that when I was born the disease was not diagnosed when Guthrie test was done. But when I was three years old, another test was done and then the disease was diagnosed. I had symptoms and was hospitalized at the age of nine months, nobody knew what was going on. It happened three years ago, I was hospitalized for a long time. When I was six years old, but only after my sister was born because she was diagnosed with the disease and then I had to take the test."* (DSC-B).

How do they perceive themselves as compared to their peers

Asked about how they perceived themselves compared to other adolescents not affected by SCD, most respondents (83.3%) said they did not feel different from their peers, while the others (16.7%) reported feeling different from their peers because of the limitations imposed by the disease, as follows.

Feels different: *"Sometimes I cannot run long because of my leg, it hurts. They(friends)say that I'm not very different from them, but I am more limited. While they keep running, I have to stop sometimes... But I don't think the others will notice that I'm tired. After too much running, I feel shortness of breath and feel sick..it's weird... I think I am short... After the vacation, I noticed that everyone grew, but I am the same size"* (DSC-A).

Interference of sickle cell disease in the daily life

Although they did not perceive themselves as different from the others because of the disease, the respondents recognize that SCD interferes with their daily lives, especially (62.5%) physically, with a potential limiting factor.

Physically: *"Sometimes I have pain episodes. And there are times that I feel very sick and end up with a very strong headache. When I run, I feel pain in the leg and below the rib. When I run, I don't feel much tired... when I run long, I feel shortness of breath. During physical education classes, I feel sometimes tired and my teacher thinks I am making a fuss about it... it is because he does not yet know about my condition... In my appointment today my doctor will write a letter explaining the disease, which will be handed to the teacher. Sometimes I feel a very strong pain... I can't run long, if I run long my foot hurts"*

and I feel a little tired. Yes, that's right: the physical activities are the most affected by SCD" (DSC-A).

Socially: *"Interferes a little, because I miss classes, cannot see my friends and this is not good... I stay quiet in my corner... I lie and my mother gives me medicine... and then I am admitted to the hospital"* (DSC-B).

Routine medications and appointments

In order to obtain the responses to the therapies administered in the outpatient clinic, the setting of this research, the discourses of the participants who described the therapies, the medications administered and the frequency of the consultations were analyzed. Two categories were defined as synthetic discourses: routine medications and appointments, with a sharing power of 52.2% and 47.8%, respectively.

Consultations: *"I come once a month... every five days.. every two months... every three months or four months...that depends, I guess. I come to the appointment and if I am well, I can go away; if I'm not well.... then I stay here"* (DSC-A).

Medication: *"I take a medicine prescribed for people with SCD, hydroxyurea, folic acid, captopril and digoxin. Hydroxyurea is for SCD; I take captopril to lower blood pressure, and I take dipyrone and paracetamol for pain relief. Sometimes I feel pain... yesterday I had a headache, then I took the medicine and the pain was gone. I come to this outpatient care clinic to make the blood transfusion. The health professionals say that this is necessary to "remove the cells with anemia"* (DSC-B).

Perception of care in the outpatient care clinic

In this discourse, the adolescents were asked about how they perceived care in the outpatient unit and if they had suggestions for improvement of the services in the outpatient care clinic. Most respondents (91.7%) expressed satisfaction with the services provided in the outpatient clinic, and only one participant expressed dissatisfaction related to the disease, not the outpatient care clinic.

Expresses satisfaction: *"No, 'everything is fine. That's great" I like to come here because of the school. I have no suggestions for improvement"* (DSC-A).

Dissatisfied with the disease: *"More or less... I'm not very fond of it because besides not liking to get up early, I don't like to think that I have this disease. I don't feel bad about having the disease... I just don't like to come here"* (DSC-B).

DISCUSSION

Despite the prevalence of SCD among women, there is no evidence of a relationship between gender and SCD risk⁽⁹⁾. A study conducted in Uberaba, in 2008, identified that 52.4% people with SCD were female⁽⁵⁾. This prevalence can be related to the slight predominance of women (4.2%) in the Brazilian population⁽¹⁰⁾.

The fact that most participants were 12 years old and that almost 70% were diagnosed through Newborn Screening Program highlights the positive outcome of the National Newborn Screening Program in 2001⁽¹¹⁾ and regulates the actions targeted to early diagnosis and treatment of SCD. However, in four cases, hospital admission was required for diagnosis of the disease, a situation that may indicate the existence of maternity hospitals where neonatal screening is not regulated. In Paraná, with the detection of SCD in neonatal screening, the newborn is referred to a reference service for neonatal screening, the FEPE (*Centro de Pesquisas da Fundação Ecumênica de Proteção ao Excepcional*) where diagnosis is confirmed and the patient is referred to a specialized service for clinical treatment⁽¹²⁾.

A qualitative study conducted in Pernambuco with escorts of patients with SCD indicated that care is centered on the mother. The disease impacts the whole family affecting the relationships of the members. The study also found that there is an entrenched cultural notion that household duties are women's responsibility, while men perform paid work⁽¹³⁾. Therefore, even women who work outside the home give up their careers to care for a family member with chronic disease.

In the present study, 50% of the participants identified themselves as black and 25% as brown. Paraná has 18 municipalities with 34 surviving communities of slave descendants, and 10 traditional black communities. Paraná was also the first Brazilian state to include the hemoglobin electrophoresis test in prenatal screening, in routine examinations of pregnant women, especially for detecting SCD⁽¹⁴⁾. A retrospective study with secondary data, in Uberaba, identified the variable skin color in only 7.8% of the participants. Of these, 4.9% were black and 1.9% were brown⁽⁵⁾. The close relationship between ethnicity and SCD justifies the need to gather accurate information on the patients' ethnic origin.

The researchers also asked the adolescents about their knowledge of the disease and found

that half of the respondents had some knowledge of sickle cell disease, even if partial or insufficient. The fact that half of the respondents reported lack of knowledge of the disease is disturbing, since such knowledge is necessary for prevention, self-care and compliance with the treatment. An analysis of the deaths of patients with SCD in Minas Gerais indicated that 40% of them occurred around 24 hours after the onset of symptoms. Along with awareness of the period of highest risk for complications and mortality, the authors mentioned continuing training of the health care staff and health education for the patients' family members as essential for the detection and prevention of complications and death⁽¹⁵⁾.

Most adolescents interviewed in the survey (83.3%) said they did not perceive themselves as different from their peers because of SCD, a genetic and chronic disease that affects the whole family since the birth of the affected individual. The collective subject discourses of the participants indicate that the disease causes conflict situations related to the pain and limitations of the disease itself and growth retardation. Regarding physical growth disorders, iron overload can occur due to the recurring transfusions, which can potentially generate endocrine disturbances and growth retardation⁽¹⁶⁾.

Painful episodes may last for 2 hours or longer, occasionally affecting arms, legs, back, abdomen, chest and head. They are strongly associated with a poor quality of life⁽¹⁷⁾ and are the main causes of hospitalization^(17,9). The symptoms identified by family members at the beginning of the disease were fever, paleness and vomiting, and pain episodes were recurrent in nearly 60% of the children⁽¹⁵⁾. This situation affects self-esteem and socialization because of the frequent hospitalizations of the patients⁽¹⁸⁾. The continued use of hydroxyurea is very important as it favors the increase in hemoglobin to fight hemolytic anemia. However, the only curative treatment is stem cell transplantation⁽¹⁹⁾. In this regard, we emphasize that there are gaps in the identification of the complications of SCD by health professional and family members⁽¹⁵⁾, which makes it difficult to treat the disease in a timely manner.

Preventive measures are needed to control pain and treat infections, with gains in life expectancy and quality. Thus, attendance to medical appointments at reference services and monitoring impact sociability and schooling. Learning about the experiences of individuals with SCD allows the organization and prioritization of nursing care, as well as actions aimed to help

promote family balance. Encouraging self-care is important in adaptation and means zeal with the body, favoring self-observation and observation of body signs⁽²⁰⁾.

The adolescents expressed satisfaction with the services provided by the outpatient care clinic, which probably reflected in the quality of life after the beginning of the treatment. User satisfaction favors accessibility, the formation of bonds and openness to the work performed by the multidisciplinary team that provides care to SCD patients⁽¹³⁾.

The dissatisfaction expressed by some respondents was related to the disease, which is common among chronic patients, because the process of accepting a chronic disease and its limitations is challenging and generates mixed feelings. Therefore, we highlight the importance of psychotherapy support to help the patients accept their condition⁽²¹⁾ and coping strategies.

FINAL CONSIDERATIONS

The present research that involved a limited number of participants and was restricted to the reference service indicates the representation of the chronic disease and its impact on the patients' lives. The findings may assist in establishing possibilities and priorities for comprehensive health care to sickle cell patients.

(Re) thinking health education actions focused on the particularities of each family and adolescent patient is recommended to successfully minimize the impacts of the chronic disease and promote quality of life. Adolescence involves questions that need specific information, support and clarifications regarding the impact of SCD on their growth, development and socialization.

More reference services for hemoglobinopathies are needed in the state of Paraná, since almost half of the users of the outpatient care clinic, the setting of this study, came from other municipalities. The frequent displacements have a negative impact on schooling and probably on the professional activities of their caregivers.

We recommend that the professionals involved in the assistance to adolescents with SCD receive continuing training in their workplaces. Further studies in different settings and with varied populations (e.g. family members, health professionals and specialized service managers) could contribute to a better understanding of the impact of sickle cell disease.

REFERENCES

1. Piel FB, Hay SI, Gupta S, Weatherall DJ, Williams TN. Global burden of sickle cell anaemia in children under five, 2010–2050: Modelling based on demographics, excess mortality, and interventions. *PLOS Med.* [Internet] 2013; 10(7) [acesso em 07 jul 2015]. Disponível: [10.1371/journal.pmed.1001484](http://dx.doi.org/10.1371/journal.pmed.1001484).
2. Piel FB, Tatem AJ, Huang Z, Gupta S, Williams TN, Weatherall DJ. Global migration and the changing distribution of sickle hemoglobin: a quantitative study of temporal trends between 1960 and 2000. *Lancet*. [Internet] 2014; 2. [acesso em 07 jul 2015]. Disponível: [http://dx.doi.org/10.1016/S2214-109X\(13\)70150-5](http://dx.doi.org/10.1016/S2214-109X(13)70150-5).
3. Santos JP, Gomes Neto M. Sociodemographic aspects and quality of life of patients with sickle cell anemia. *Rev. Bras. Hematol. Hemoter.* [Internet] 2013; 35(4). [acesso em 07 jul 2015]. Disponível: <http://dx.doi.org/10.5581/1516-8484.20130093>.
4. Ansong D, Akoto AO, Ocloo D, Ohene-Frempong K. Sickle cell disease: Management options and challenges in developing countries. *Mediterr J Hematol Infect Dis.* [Internet] 2013; 5(1). [acesso em 07 jul 2015]. Disponível: [doi:10.4084/MJHID.2013.062](http://dx.doi.org/10.4084/MJHID.2013.062).
5. Martins PRJ, Souza HM, Silveira TB. Morbidity-mortality in sickle cell disease. *Rev. Bras. Hematol. Hemoter.* [Internet] 2010; 32(5). [acesso em 07 jul 2015]. Disponível: <http://dx.doi.org/10.1590/S1516-84842010000500010>.
6. Lefèvre F, Lefèvre AMC. The Collective Subject that speaks. *Interface.* 2006; 10(20):517-24.
7. Jodelet D. Ponto de vista: sobre o movimento das representações sociais na comunidade científica brasileira. *Temas Psicol.* 2011, vol.19, n.1, pp. 19-26. ISSN 1413-389X.
8. Tong A, Sainsbury P, Graig J. Consolidated criteria for reposrtng qualitative research (COREQ): a 32-item check list for interviews and focus group. *Inter.Journ. Qual. Health Care.* [Internet] 2007; 19(6). [acesso em 07 jul 2015]. Disponível: DOI: <http://dx.doi.org/10.1093/intqhc/mzm042>.
9. Santos PND, Freire MHS, Zanlorenzi GB, Pianovski MA, Denardi VFAM. Anemia falciforme: caracterização dos pacientes atendidos em um ambulatório de referência. *Cogitare Enferm.* [Internet] 2014; 19(4). Disponível: <http://dx.doi.org/10.5380/ce.v19i4.36657>.
10. Instituto Brasileiro de Geografia e Estatística (IBGE). Censo demográfico e contagem da população 2010. [Internet] [acesso em 07 jul 2015]. Disponível: <http://censo2010.ibge.gov.br/resultados>.
11. Brasil. Ministério da Saúde. Secretaria de Atenção à Saúde. Departamento de Atenção Especializada. Doença falciforme: condutas básicas para o tratamento. Brasília: Ministério da Saúde, 2012. [acesso em 07 jul 2015]. Disponível: http://bvsms.saude.gov.br/bvs/publicacoes/doenca_falciforme_condutas_basicas.pdf.
12. Federação Espírita do Paraná. Teste do Pezinho, 2014. [Internet] [acesso em 07 jul 2015]. Disponível: <http://www.fepe.org.br/triagem.html>.
13. Guimarães TMR, Miranda WL, Tavares MMF. The day-to-day life of families with children and adolescents with sickle cell anemia. *Rev. Bras. Hematol. Hemoter.* [Internet] 2009; 31(1). [acesso em 07 jul 2015]. Disponível: <http://dx.doi.org/10.1590/S1516-848420090005000002>.
14. Secretaria Estadual de Saúde do Paraná. Boletim Informativo Semanal – e-Saúde. Paraná [Internet] 2014 [acesso em 07 jul 2015] Disponível: <http://www.sesa.pr.gov.br/modules/conteudo/conteudo.php?conteudo=2867>
15. Sabarense AP, Lima GO, Silva LML, Viana MB. Characterization of mortality in children with sickle cell disease diagnosed through the Newborn Screening Program. *J Pediatr.* [Internet] 2015; 91. [acesso em 07 jul 2015]. Disponível: [doi:10.1016/j.jpeds.2014.08.006](http://dx.doi.org/10.1016/j.jpeds.2014.08.006).
16. Felix AA, Souza HM, Ribeiro SBF. Epidemiologic and social aspects of sickle cell disease. *Rev. Bras. Hematol. Hemoter.* [Internet] 2010; 32(3). [acesso em 07 jul 2015]. Disponível: [doi:10.1016/j.jpeds.2014.08.006](http://dx.doi.org/10.1016/j.jpeds.2014.08.006).
17. Makani J, Ofori-Acquah SF, Nnodu O, Wonkam A, Ohene-Frempong K. Sickle cell disease: new opportunities and challenges in Africa. *The Scientific World Journal.* [Internet] 2013, 193252. [acesso em 07 jul 2015]. Disponível: <http://dx.doi.org/10.1155/2013/193252>.
18. Dias TL, Oliveira CGT, Enumo SRF, Paula KMP. Pain the daily life of caregivers and children with sickle cell anemia. *Psicol. USP.*[Internet] 2013; 24(3). [acesso em 07 jul 2015]. Disponível: <http://dx.doi.org/10.1590/S0103-65642013000300003>
19. Editorial. Boosting services for people with sickle-cell disease. [Internet] 2010; 376. [Acesso em 07 jul 2015]. Disponível: DOI: [http://dx.doi.org/10.1016/S0140-6736\(13\)60030-6](http://dx.doi.org/10.1016/S0140-6736(13)60030-6).
20. Cordeiro RC, Ferreira SL, Santos ACC. Experiências do adoecimento de pessoas com anemia falciforme e estratégias de autocuidado. *Acta paul. enferm.* [Internet] 2014; 27(6). [acesso em 07 jul 2015]. Disponível: <http://dx.doi.org/10.1590/1982-0194201400082>.
21. Savassi LCM. Patient Satisfaction and Self-perceived Health in Primary Care. *R. Bras. Med. Fam. e Comum.* [Internet]. 2010; 5(17). [acesso em 7 jul 2015]. Disponível: DOI: [http://dx.doi.org/10.5712/rbmfc5\(17\)135](http://dx.doi.org/10.5712/rbmfc5(17)135).