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Barriers experienced in self-care practice by young people with sickle cell disease

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ABSTRACT

Objective: To identify barriers to the self-care practice of young people with sickle cell disease.
Method: This qualitative study was conducted with 17 individuals with sickle cell disease aged between 13 and 24 years in Belo Horizonte, MG, Brazil in March and April 2017. An interview investigated the barriers to self-care practice and the feelings associated with sickle cell disease. Data were transcribed and analyzed according to Bardin's perspective using the following steps: (1) pre-analysis, (2) exploration of the material, and (3) treatment of the results (inference and interpretation).
Results: Five thematic categories emerged: (1) feelings: anger, sadness, and fear; (2) bullying and stigmatization: challenges regarding walking, speaking, or behaving, as well as patient labels; (3) cognitive factors: doubts related to medication, hydration, heredity and maternity; (4) medication compliance: fear of the side effects suffered and anger triggered by the obligation to use the medication; (5) family issues: complaints of not earning the mothers' trust to live independently.
Conclusion: The barriers to self-care in young people with sickle cell disease indicate difficulties related to emotional, behavioral, and environmental aspects. Understanding these factors will favor a better adaptation of youths to the context of sickle cell disease.

Introduction

The transition period between childhood and adulthood, called adolescence, represents a challenge in the management of different chronic conditions. This problem is even harder for young people with sickle cell disease as it requires complex care. The need of learning to take care of oneself with autonomy, to be responsible for health choices, and to adapt to illness are some of the difficulties faced by the youths.[1]

To take care of yourself, referred to as self-care, comprises a process in which young people with sickle cell disease perform activities to improve their health and well-being, requiring attitudes compatible with medical guidelines associated with efforts to minimize disease-related complications. These actions include, among other things, the correct use of

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medication, hydration, and care about extreme temperatures and of high impact activities.[2,3]

Nevertheless, adherence to self-care practices might be permeated by barriers that demand an understanding of their multifactorial aspects and the characteristics to which they may be associated.[4] The literature indicates that barriers pervade the behavioral, sociocultural, and psychosocial domains. As an example, cultural beliefs about disease, socioeconomic status, presence or absence of social support, and past negative experiences with medical teams play key roles in adherence to self-care.[5,6]

Seeking to identify and understand the barriers experienced by young people with sickle cell disease is essential to plan and systematize better health care practices to improve adherence to self-care and promote the well-being of this population.

Objective

In this perspective, this study aimed to identify barriers to self-care practice in young people with sickle cell disease.

Method

This is a descriptive and exploratory qualitative study conducted with 17 young people with sickle cell disease treated in the Outpatient's Clinic of the Fundação Centro de Hematologia e Hemoterapia do Estado de Minas Gerais in Belo Horizonte (Hemominas), MG, Brazil, part of the Centro de Educação e Apoio para Hemoglobinopatias de Minas Gerais (CEHMOB) during March and April 2017.

The inclusion criteria were individuals aged between 13 and 24 years diagnosed with sickle cell disease [hemoglobin (Hb) SS, Hb SC, and Hb Sβ-Thalassemia] and able to understand and answer questions. Data collection occurred through semi-structured face-to-face interviews conducted in a reserved room at the Hemocentro, without noise and adequate for dialog. The research reached the saturation criteria proposed for qualitative research by the 17th interview. All individuals accepted to participate in the research.

Each interview, carried out by the authors of this article, lasted for an average of 20 min. The interviews were recorded after the participants' consent and guided by the following questions: (a) for you, what is it like living with sickle cell disease? (b) tell me about the difficulties you face to take care of yourself and your health and (c) how do you feel about having to take care of yourself and your health? The questions were read in colloquial Portuguese, familiar to the context of each subject. All interviews were later transcribed using the ELAN software.

The contents of the interviews were organized after the complete transcription of the recordings, preserving their originality. For data analysis, these dialogs were systematized and categorized to compose a database considering recurrent opinions, dissent and consensus about the barriers related to self-care. Later, data processing and interpretation took place based on the content analysis technique proposed by Bardin that consisted in the following stages: (1) pre-analysis, partially guided reading of the material so that the researcher could become familiar with the expressed content, (2) exploration of the material, requiring several readings and reinterpretations, during which the material was organized so that the initial ideas were systematized and (3) treatment of the results, where all the material was separated into record units regarding each topic and category (inference and interpretation). It is also important to clarify that, to guarantee the anonymity of participants, the statements were codified by letters (I) and numbers (1–17). Reading and interpretation of the content identified five thematic categories: feelings, bullying and stigmatization, cognitive factors, medication compliance, and family issues.

To describe the profile of participants according to sociodemographic variables, a structured questionnaire containing information about age, gender, place of origin, racial background, education, and type of sickle cell disease was applied.

The study respected the formal requirements established in the national and international norms regulating research involving human beings and every research participant signed the free and informed consent form after it was approved by the Research Ethics Committees of the Universidade Federal de Minas Gerais (UFMG) and Hemominas (# 58078316.0.0000.5149).

Results

The sample consisted of 17 people with a diagnosis of sickle cell disease with a mean age of 17.8 years. Regarding gender, 47.1% were male and 52.9% were female. More than one-third (35.2%) had incomplete basic education, 52.9% had incomplete high school education, and 11.7% had quit school. About the origin, 52.9% were from Belo Horizonte and the metropolitan region and 47.1% from surrounding towns. As for the racial background, 64.7% declared themselves Black and 35.2% mixed (Black and White). The monthly family income of 11.7% of the cases was less than one minimum wage, another 11.7% of the families received between one and three minimum wages, 5.8% between three and five minimum wages, and the rest did not know or chose not to disclose the family income. Regarding the hemoglobin type of sickle cell disease, 41.17% had Hb SC, 29.4% had Hb SS, 5.8% had Hb Sβ-Thalassemia, and the others were unable to inform the type.

Categorical analysis identified barriers faced by young people with sickle cell disease in respect to self-care or even to attempting to look after themselves. Based on the analyzed reports, it was possible to group and classify the barriers in five thematic categories.

Feelings

Given the chronicity of sickle cell disease, emotional reactions related to barriers to self-care were identified with special reference to the feelings of anger, sadness and fear.

Anger was mentioned by most of the youths and was associated with the presence of pain, because, if it were not for the disease, there would be no such symptom or any other complication. It was observed that some participants who reported anger due to the sickle cell disease presented resistance to

medication and self-care practice, including related to heavy physical exercise and water intake.

I5: *I feel angry because it is too bad, no one else has it [besides us]. I know that we go to the hospital and see people in a much worst situation, but it is bad to see my sister having a crisis, we both have this disease, for me it is okay, but seeing my sister having a crisis, crying with pain [. . .] it puts me down.*

Participants also reported sadness. Among the reasons, many mentioned the fact of having a chronic illness and experiencing consequent limitations of pain crises, frequent hospitalizations, and concerns about the possibility of long-term complications.

I6: *It is very annoying, you know? It is bad to always have pain. . . Oh, I am going to cry (cry). I cannot speak [. . .].*

I7: *When it [the disease] got much worse, I felt useless at home. Because I started doing something then I felt dizzy, weak, sometimes I fainted, I felt a lot of pain.*

Fear was presented by young people as one of the barriers to self-care, as it impacts on future perspectives such as maintaining employment and the need to deal with a chronic illness, taking medications for life, and not being able to manage your health.

I7: *For me, it is like a thorn in the side because it is very difficult to live with this disease, it affects many things in our life, as in our day-to-day, I do not know if I'll get a job, work, get married, live, you know? It disturbs me, I do not know if I can take care of myself.*

Furthermore, the impact on choices, such as motherhood, creates apprehension about the consequences.

I4: *I wanted to be a mother, I want to be a mother. But with this disease, I do not know, I am afraid. The doctor made me afraid.*

Bullying and stigmatization

Bullying concerning how they walk, speak, or behave was cited by the study participants as a situation that hinders the self-care practice. Expressions like 'yellow eyes', 'butter' (due to the difficulty of carrying out heavy work), and 'soft bone' (given the pain related to the disease) were listed by the youths as faced in daily school life.

As for stigmatization, some reported not feeling sick, but being treated as sick and thus not feeling strong enough to overcome the adversities of sickle cell disease. Many people feel different from their peers and other family members and cannot deal with it if they are assigned a stereotype of abnormality and inadequacy.

I13: *People call me sick, they treat me differently. I feel that they do not like me, and it is because of the disease [. . .] I feel like I am different from other people, I feel it [. . .]*

I15: *At home, I cannot do anything, I am considered incapable [. . .] and I end up feeling incapable [. . .]*

Cognitive factors

The youths demonstrated not understanding sickle cell disease, unable to explain why they had it, why the treatment should be rigorously followed, and showed a lack of knowledge. Doubts related to medication, hydration, heredity, and maternity were mentioned by the participants when asked about the barriers they faced to take care of themselves.

I15: *I don't know why I have this disease, or why I feel this [pain]. Sometimes, I need to come here and do a transfusion, but why, I don't know. I think that it is to avoid my pain.*

I4: *And why I can't be a mother? It's because of my disease, I just know it.*

Medication compliance

Regarding medications, some participants stated that they did not adhere to the use of medications because they did not understand their benefits or their importance to control sickle cell disease. Most of the individuals feared the side effects they could experience, as well as the anger triggered by the obligation to use the medication, often from a demand of parents/guardians or health care professionals. Some said that when they felt obliged to take the medications they developed resistance and preferred to stop it, even though this represented a significant loss in disease control.

I1: *I have it [resistance to medication] because I do not feel like taking it [the medication], because I feel obliged to take the medicine. And if I do not want to take it [the medication], I do not have to take it.*

Family issues

This category refers to the relationship between the participants with sickle cell disease and their mothers, which may be determinant for developing the self-care practice. Some have expressed conflict situations with their mothers, holding them responsible for having the disease and for not allowing their autonomy in respect to care. Furthermore, they complained about not being able to earn their mothers' trust to go out alone, to get into relationships with others, or simply to live independently.

I3: *[. . .] My mother suffocates me [. . .] she will not let me do anything, I know she is afraid that I'll be taken ill out on the street, but she must understand that I am already 22 [. . .] I am already a woman, I have grown up, I want to discover the world [. . .]*

On the other hand, the lack of family support was also reported during the interviews, since it represents the lack of emotional support appropriate for the maintenance of care, which requires frequent visits to the outpatient clinic for hematology treatment.

Discussion

Sickle cell disease requires people to follow a rigorous care plan, focusing on the correct use of specific drugs that help control its symptoms and prevent complications that could

endanger the health and life of youths. A few years ago, sickle cell disease was considered mostly a childhood disease, and actions that could help care and positively impact the lives of people with this disease were not discussed.[7–9]

Nonetheless, with advances in the health area, such as the implementation of the Newborn Screening Program (PETN) in the state of Minas Gerais, there was a change in the late diagnosis scenario, introducing a new panorama of the disease in terms of public health.[10] In this context, the life expectancy of this population increased and along came the discussions about factors that influence adherence to self-care.[8]

Hindering factors, called barriers, prevent young people with sickle cell disease from developing adequate care and managing the condition effectively. The literature conceptualizes self-care as a routine delimited by schedules, food restriction, use of medications, tests, and hemoglobin control. Additionally, there are weather-related factors that involve protection against the sun and cold. However, at some point in life, these practices become less frequent, especially in adolescence and the transition to adulthood.[11]

In this sense, one study indicates that identifying and approaching the factors that lead to not performing self-care allows young people with sickle cell disease to improve their quality of life and reduce hospital assistance and admissions.[5] Therefore, this study demonstrated the need to go beyond conventional barriers and understand that the limitations caused by the emotional aspects of sickle cell disease are also responsible for the in-treatment results and impact on the well-being of youths.

Another study showed that when the difficulties of young people and adults with sickle cell disease are heard, accepted, and worked out, the opportunity to turn negative feelings into positive ones increases.[12] Thus, overcoming barriers depends mostly on the young people's interaction with their own feelings related to the chronic condition, which must be organized, understood, and worked out to overcome the suffering caused by the illness.[13]

The feelings category found in this study indicated an interrelationship between the occurrence of negative feelings and barriers, so that the greater the occurrence of negative feelings, the more barriers are identified in the development of self-care, which strengthens the idea of developing individualized care plans and strategies that can help these patients to manage their emotions.

Regarding sadness, the literature reports that the occurrence of this feeling may be associated with the restrictions imposed by the disease, which in youths reflects the inability to perform certain recreational activities and sports that in some cases may lead to depression and anxiety.[14,15]

The chronicity of sickle cell disease is characterized by uncertainty for the future; although young people and adults experience periods of stability, there is a constant sense of insecurity and apprehension concerning the next day.[13] The findings of this study corroborated what is reported in the literature, since the participants expressed fear about adult life, encompassing aspects such as maternity, work expectancy, and self-care.

As for medication resistance presented in the medication compliance category, the literature confirms the observed results, since young people with sickle cell disease present relationship problems and tend to have attitudes contrary to those indicated by the family or health team.[16]

Regarding stigmatization, some authors define it as a process of associating a negative aspect to a characteristic of an individual or group. The stigma experienced by these youths is one of the many obstacles to overcome to adapt to the chronic condition and adhere to self-care.[17] In this study, the stigma was associated to bullying and a feeling of social inadequacy.

In the context of sickle cell disease, bullying concerns the physical changes related to short stature, delayed sexual maturation, jaundice, and restrictions imposed by treatment, hence contributing to the appearance or aggravation of psychosomatic and behavioral problems.[15]

Knowledge about sickle cell disease is still a challenge for the population and health professionals, a condition that can result in inadequate care.[18] Furthermore, the literature highlights that a good relationship between health professionals and these patients is essential, because through this relationship young people are encouraged to participate in their care and achieve empowerment to manage their lives.[12]

Additionally, young people with sickle cell disease must deal with their own difficulties in understanding the disease and its care, determinants for the development of autonomy in the transition phase into adulthood. The present study demonstrates that this knowledge is still insufficient and endangers the health condition of youths.[1]

One study carried out in three municipalities in the state of Bahia, Brazil, aiming to know the experiences related to illness of patients with sickle cell disease and their self-care strategies, indicated that when people with this condition have sources of information about the disease and understand it, they become more open and willing to face the challenges and barriers to self-care. Moreover, the authors of this research concluded that knowledge about the disease is equally important to help understand aspects of life, realign the present, and design actions for the future.[13]

Concerning family issues, the assistance received from parents or guardians is a key factor for the motivation to health care. In a study conducted in Ohio, United States, participants reported that they are more likely to attend a clinical consultation if their parents or guardians are present.[5] On the other hand, the excess of attention becomes a barrier to self-care, as identified in this study, since the youths become dependent and feel incapable of managing their own health.

Understanding the barriers to self-care presented in this study enabled a closer look to the reality of young people with sickle cell disease and presented a panorama that needs to be better explored to encourage the maintenance of care. The findings of this study imply the need to develop educational strategies that motivate adherence to self-care by young persons with sickle cell disease and to help overcome barriers in behavioral, emotional, and environmental aspects.

Efforts at the educational, institutional, and cultural levels are necessary to develop actions that favor an ongoing care process.[19] Therefore, the authors suggest that practitioners working with sickle cell disease care should focus on developing educational interventions to teach skills that encourage the youths to face the barriers, hence achieving emotional well-being.

As demonstrated in other chronic diseases, when health professionals are prepared and proactive and create conditions of interaction with an informed and empowered youth, care becomes shared and promotes better clinical and emotional control.[20]

The characteristics of this study do not allow the results to be generalized, but rather to understand the barriers faced by young people with sickle cell disease under treatment at the Hemocentro of Belo Horizonte (Hemominas) in the context and period specific to the interviews conducted, thus not presenting limitations that would interfere with the results of this research. Nonetheless, the authors suggest the development of further investigations to deepen the understanding of this issue to better explore the problem and find solutions.

Conclusion

The barriers to self-care in young people with sickle cell disease indicate difficulties related to emotional, behavioral, and environmental aspects. It is necessary to understand them in depth in order to propose measures to promote self-care aiming at a better adaptation of the youths to the context of sickle cell disease.

Conflicts of interest

The authors declare no conflicts of interest.

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REFERENCES

1. Lebensburger JD, Bemrich-Stolz CJ, Howard TH. Barriers in transition from pediatrics to adult medicine in sickle cell anemia. J Blood Med [Internet]. 2012;3:105–12. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3460672/ [cited 22.08.17].
2. Araujo PI. O autocuidado na doença falciforme. Rev Bras Hematol Hemoter [Internet]. 2008;29(3):239–46. Available from: http://www.scielo.br/pdf/rbhh/v29n3/v29n3a10.pdf [cited 22.08.17].
3. Matthie N, Jenerette CM, McMillan S. The role of self-care in sickle cell disease. Pain Manag Nurs [Internet]. 2015;16(3):257–66. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4417084/ [cited 22.08.17].
4. Vargas EC, Cecilio SG, Brasil CL, Torres HC. Identificando barreiras e cumprimento de metas para prática de autocuidado em pessoa com diabetes tipo 2. Cogitare Enferm [Internet]. 2015;20(4):846–50. Available from: http://revistas.ufpr.br/cogitare/article/view/42572 [cited 22.08.17].
5. Crosby LE, Modi AC, Lemanek KL, Guilfoyle SM, Kalinyak KA, Mitchell MJ. Perceived barriers to clinic appointments for adolescents with sickle cell disease. J Pediatr Hematol Oncol [Internet]. 2009;31(8):571–6. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2750821 [cited 22.08.17].
6. Tanabe P, Porter J, Creary M, Kirkwood E, Miller S, Ahmed-Williams E, et al. A qualitative analysis of best self-management practices: sickle cell disease. J Natl Med Assoc [Internet]. 2010;102(11):1033–41. Available from: http://www.sciencedirect.com/science/article/pii/S0027968415307306?via%3Dihub [cited 22.08.17].
7. American. Society of Hematology. State of Sickle Cell Disease: 2016 Report. Washington: American Society of Hematology [Internet]; 2016. Available from: http://www.scdcoalition.org/pdfs/ASH%20State%20of%20Sickle%20Cell%20Disease%202016%20Report.pdf [cited 22.08.17].
8. Serjeant GR. The natural history of sickle cell disease. Cold Spring Harb Perspect Med [Internet]. 2013;3(a011783):1–11. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3784812/ [cited 22.08.17].
9. Kanter J, Jordan LB. Improving the healthcare model for management of adults with sickle cell disease in the PPACA era. J Hematol Transfus [Internet]. 2015;3(1):1037. Available from: https://www.jscimedcentral.com/Hematology/hematology-3-1037.pdf [cited 22.08.17].
10. Ministério da Saúde (Brasil), Secretaria de Atenção à Saúde, Departamento de Atenção Especializada. Doença falciforme: atenção integral à saúde das mulheres. 1a edição Brasília; 2015. Available from: http://bvsms.saude.gov.br/bvs/publicacoes/doenca_falciforme_atencao_integral_saude_mulher.pdf [cited 22.08.17].
11. Hirschberg M. Living with chronic illness: an investigation of its impact on social participation. Reinvention [Internet]. 2012;5(1.). Available from: http://www.warwick.ac.uk/reinventionjournal/issues/volume5issue1/hirschberg [cited 22.08.17].
12. Wright K, Adeosun O. Barriers to effective pain management in sickle cell disease. Br J Nurs [Internet]. 2009;18(3):158–61. Available from http://www.magonlinelibrary.com/doi/10.12968/bjon.2009.18.3.39043 [cited 22.08.17].
13. Cordeiro RC, Ferreira SL, Santos AC. Experiences of illness among individuals with sickle cell anemia and self-care strategies.
Acta Paul Enferm [Internet]. 2014;27(6):499–504. Available from: http://www.scielo.br/pdf/ape/v27n6/en_1982-0194-ape-027-006-0499.pdf [cited 22.08.17].
14. Hijmans CT, Grootenhuis MA, Oosterlaan JL, Heijboer H, Peters M, Fijnvandraat K. Behavioral end emotional problems in children with sickle cell disease and healthy siblings: multiple informants, multiple measures. Pediatric Blood Cancer [Internet]. 2009;53:1277–83. Available from: https://www.ncbi.nlm.nih.gov/pubmed/19731327 [cited 22.08.17].
15. Lorencine GR, Paula KM. Perfil Comportamental de Crianças com Anemia Falciforme. Temas Psicol [Internet]. 2015;23(2):269–80. Available from: http://pepsic.bvsalud.org/scielo.php?script=sci_arttext&pid=S1413-389X2015000200003 [cited 22.08.17].
16. Benton TD, Boyd R, Ifeagwu JA, Feldtmose E, Smith-Whitley K. Psychiatric diagnosis in adolescents with sickle cell disease: a preliminary report. Curr Psychiatry Rep [Internet]. 2011;13(2):111–5. Available from:

https://www.ncbi.nlm.nih.gov/pubmed/21312010 [cited 22.08.17].
17. Jenerette CM, Brewer C. Health-related stigma in young adults with sickle cell disease. J Natl Med Assoc [Internet]. 2011;102(11):1050–5. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3119340/ [cited 22.08.17].
18. Clay OJ, Telfair J. Evaluation of a disease-specific self efficacy instrument in adolescents with sickle cell disease and its relationship to adjustment. Child Neuropsychol [Internet]. 2007;13(2):188–203. Available from: https://www.ncbi.nlm.nih.gov/pubmed/17364574 [cited 22.08.17].
19. Matthie N, Jenerette C, Mcmillan S. The role of self-care in sickle cell disease. Pain Manag Nurs [Internet]. 2015;16(3):257–66. Available from: https://www.ncbi.nlm.nih.gov/pubmed/25439112 [cited 22.08.17].
20. Molter BL, Abrahamson K. Self-efficacy, transition, and patient outcomes in the sickle cell disease population. Pain Manag Nurs [Internet] 2015. 2015;16(3):418–24. Available from: https://www.ncbi.nlm.nih.gov/pubmed/25047808 [cited 22.08.17].