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ORIGINAL ARTICLE

STUDY OF NON-ADHERENCE TO THE TREATMENT OF SICKLE CELL DISEASE: A FAMILY CASE STUDY

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ABSTRACT
Objective: To gain insight on the aspects that impact non-adherence to sickle cell disease treatment in a family with several sick members.
Method: Single case study of four siblings with sickle cell disease monitored at a Referral Center in the state of Bahia, Brazil. Data was collected with the use of a questionnaire, open interviews and themed story-drawing.
Results: The following themes emerged: Prejudice and discrimination promote self-isolation; Transportation problems prevent patients to get to health services; The health unit is a space that amplifies doubts and uncertainty; Hopelessness leads to treatment withdrawal.
Conclusion: Social assistance, emotional, instrumental and educational support are essential to reduce the vulnerability of families experiencing sickle cell disease. The contents addressed in health facilities must include perspectives on positive experiences, encouraging hope and the construction of meanings during life in the process of chronic illness, in order to increase treatment adherence.

DESCRIPTORS: Pharmacological treatment; Patient cooperation; Chronic disease; Prejudice; Social isolation.

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ESTUDO DA NÃO ADESÃO AO TRATAMENTO DA DOENÇA FALCIFORME: O CASO DE UMA FAMÍLIA

RESUMO

Objetivo: compreender os aspectos que influenciam a não adesão ao tratamento da doença falciforme em uma família com vários membros adoecidos.

Método: estudo de caso único realizado com quatro irmãos com doença falciforme acompanhados em um Centro de Referência no estado da Bahia, Brasil. Os dados foram colhidos por questionário, entrevista aberta e desenho-estória com tema.

Resultados: os temas emergidos foram: Preconceito e discriminação promovem o autoisolamento; Dificuldade para deslocamento impede as idas ao serviço; A unidade de saúde é um espaço que amplia dúvidas e incertezas; Desesperança leva à desistência do tratamento.

Conclusão: amparo social, suporte emocional, instrumental e educacional são essenciais para reduzir a vulnerabilidade da família que vivencia a doença falciforme. Conteúdos abordados nas unidades de saúde necessitam incluir perspectivas de experiências positivas, fomentando esperança e construção de sentidos para a vida no processo de adoecimento crônico, a fim de aumentar a adesão ao tratamento.

DESCRITORES: Tratamento farmacológico; Cooperação do paciente; Doença crônica; Preconceito; Isolamento social.

ESTUDIO DE LA NO ADHESIÓN AL TRATAMIENTO DE LA ENFERMEDAD FALCIFORME: CASO DE UNA FAMILIA

RESUMEN

Objetivo: comprender los aspectos que influyen en la no adhesión al tratamiento de la enfermedad falciforme en una familia con varios miembros enfermos.

Método: estudio de caso único que se realizó con cuatro hermanos con enfermedad falciforme asistidos en un Centro de Referencia en el estado de Bahia, Brasil. Se obtuvieron los datos por medio de cuestionario, entrevista abierta y dibujo historia con tema.

Resultados: los temas que resultaron del proceso fueron: Prejuicio y discriminación promueven el auto aislamiento; Dificultad para desplazarse imposibilita ir al trabajo; La unidad de salud es un espacio que aumenta dudas e incertidumbres; Desesperanza lleva a desistimiento del tratamiento.

Conclusión: ayuda social, apoyo emocional, instrumental y educacional son esenciales para reducir la vulnerabilidad de la familia que vive la enfermedad falciforme. Es necesario añadir perspectivas de experiencias positivas en los contenidos que se abordaron en las unidades de salud, fomentando esperanza y construcción de sentidos para la vida en el proceso crónico de la enfermedad, para aumentar la adhesión al tratamiento.

DESCRIPTORES: Tratamiento farmacológico; Cooperación del paciente; Enfermedad crónica; Prejuicio; Aislamiento social.

INTRODUCTION

Sickle cell disease (SCD) is a genetic disorder that causes a debilitating systemic syndrome. It is characterized by chronic anemia, acute painful episodes, infarction, progressive organ damage and consequently a significant reduction in life expectancy(1-2).

People with sickle cell disease experience serious complications such as infections, stroke, acute chest syndrome, priapism, leg ulcers, among others (3). Therefore, the disease must be continuously monitored, and the development of habits and attitudes that promote self-care is necessary (4).

Adherence to treatment implies following correctly the regimen prescribed by health professionals. However, this is not so simple, since this attitude is associated with multiple social, behavioral, cultural and aspects, as well as with patient/family-health professional relationship aspects (5).

Non-adherence means discontinuing the prescribed care without the advice of a health professional, either by failing to observe on a timely basis recommendations of taking medicines, receiving vaccines and adopting other protective measures, or by interrupting the prescribed therapeutic plan(6). Therefore, non-adherence may be the expression of expected results that have not been achieved, may lead to unnecessary repeat testing, changes in regimens and additional expenses(5).

Treatment adherence is essential for disease control, damage prevention and the success of the proposed therapy (7). Non-adherence may lead to premature complications, increase the number of acute seizures, require more hospitalizations, compromise the quality of life and reduce the life expectancy of the sick (6-7).

In view of the aforementioned, the present study aimed to gain insight on the aspects that impact non-adherence to sickle cell disease treatment in a family with several sick members.

METHOD

Case study, qualitative research, developed at the Referral Center for Individuals with Sickle Cell Disease in the state of Bahia, Brazil. The identification of the participants was obtained from the records of those who did not attend the appointments at the Referral Center.

The methodological-theoretical perspective of single case study, proposed by Yin (8), which is characterized by the exploration of a limited system through detailed analysis and systematic description in order to understand the specific situation of a small group. Definition of the theme/ research problem; Single case definition; Description of the theoretical propositions; Preparation of the protocol of the case study; Data collection using multiple sources of evidence; Analysis and interpretation of results; Elaboration of the report(8).

A family case study design was selected when the authors identified a case of five members of the same family group who had sickle cell disease. All of them had missed appointments at the Referral Center.

The inclusion criteria were be diagnosed with sickle cell disease and belong to the family whose members failed to attend the medical consultations in the last six months. No exclusion criteria were established. Of the five individuals who met the inclusion criteria, only one did not participate in the study because he/she was admitted to a hospital in another municipality.

Contact with the participants was made through previously scheduled home visits, with the aim of establishing a bond with them.

Data was collected between May 2016 and July 2016 at the participants' homes, individually, in a restricted environment (living room), to ensure the privacy of the subjects. Two individual meetings were held with the participants: in the first meeting, the socioeconomic and demographic questionnaire (gender, age, religion, origin, occupational status, ethnicity/color and disease complications), and themed story-drawing procedure; Open interview was used in the second meeting.

The use of the projective technique of thematic story-drawings (PDE-T) is a type of imagistic communication that favors the apprehension of statements associated with events, facts and stories that are updated in the figure (drawing) (9). A4 sulfite paper and colored pencils were made available to the participants for the drawings. Then they were told to create a drawing depicting the reasons why a person with sickle cell disease DF would not adhere to treatment. When the drawings were completed, the participants were told to observe their drawings and create a story with a beginning, middle and end. Finally, the participants were asked to read their own stories and put titles on them.

For the interviews, a guide consisting of questions related to non-adherence to the treatment and the importance of treatment for patients with SD. The interviews were recorded in MP3 and lasted in average 30 to 50 minutes. Subsequently, the drawings were scanned and the stories and interviews were fully transcribed by the first author.

All empirical material obtained through the story-drawings was submitted to the analysis for such procedures (9), according to the following sequence: systematic observation of the drawings; selection of designs by graphic similarities and/or similarities among the themes; fluctuating reading of the thematic units of the stories: perspective and classification; analysis and interpretation of thematic contents grouped by categories; analysis and interpretation of drawings through graphics. The material collected in the interviews was submitted to thematic content analysis systematized in stages: pre-analysis; exploration of the material; treatment of results; inference and interpretation (10).

Themes derived from both techniques (story-drawings and interviews) were compared, classified and inserted in four categories: Prejudice and discrimination promote self-isolation; Financial difficulty and interruption of drug supply discourage patients' visits to the referral center; the health unit is a space that amplifies doubts and uncertainty, and Hopelessness leads to treatment withdrawal.

When the analyzes were concluded, the results of the study were shared with the multidisciplinary team who continued to monitor, through home visits, the implementation of an intervention plan for the family who participated in the study. However, in this article, only data obtained at home visits was used for situational diagnosis. To preserve the participants' anonymity, their testimonies were identified by the initials INT followed by the interview order number.

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RESULTS

Four members of the same family participated in this study, as follows: three women and one man, aged 20-29 years and who lived in the same household. Regarding race /color, all declared themselves black. Regarding marital status, all were single. As for their educational level, three participants had incomplete secondary education and one had completed secondary education. Regarding occupation, all four respondents were unemployed. As for religion, all were Protestant. Regarding the complications of SCD, pain crises (four) and leg ulcer (one) predominated. All participants were diagnosed late for SCD and accessed emergency services in the last 12 months.

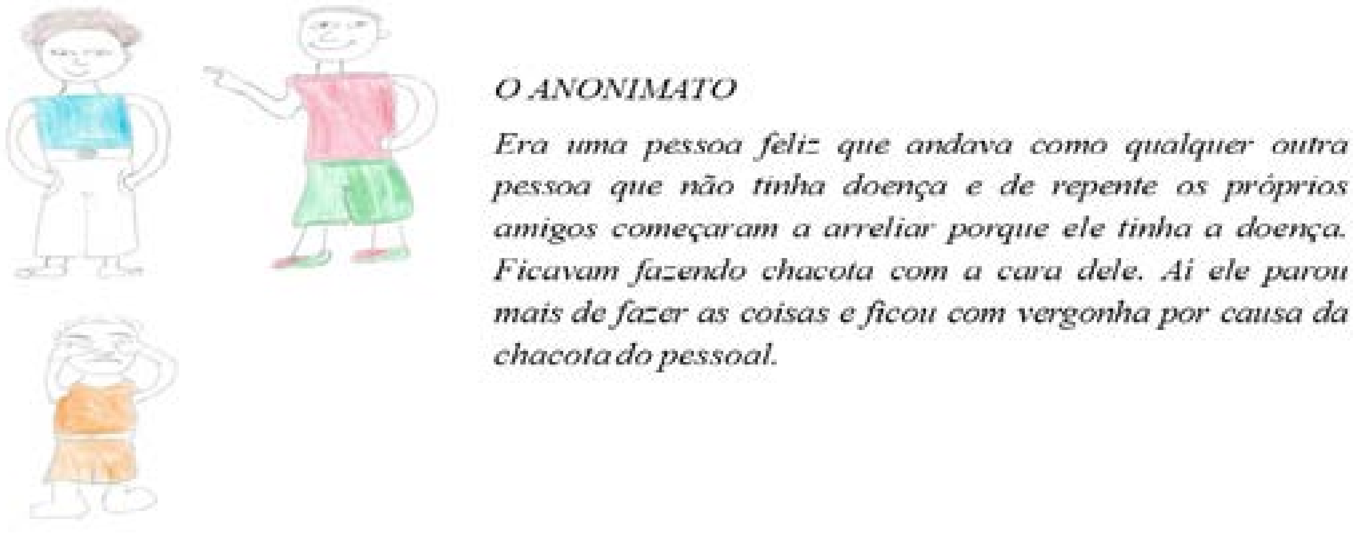
Prejudice and discrimination promote self-isolation

The prejudice and discrimination suffered by individuals with SCD contribute to the emergence of feelings of shame and withdrawal from situations that require interaction due to fear of prejudiced attitudes, which leads to social isolation (see story-drawing 1). The following statements corroborate the aforementioned:

And another thing [that results in treatment withdrawal] is the prejudice faced, as most people who face prejudice and have sickle cell disease are black, and because of the ethnicity they are discriminated. Some people think SCD is contagious; they say: "Oh, I won't get too close because I can get his disease."(INT. 2).

Many people are ashamed to say that they have [the disease] (INT 1)

Fearing that other people will learn that they have a disease, the sick individuals do not go to the appointments at the health facility, to avoid being seen there by others and referred to or treated in a derogatory manner, as shown in Figure 1.



O ANONIMATO

Era uma pessoa feliz que andava como qualquer outra pessoa que não tinha doença e de repente os próprios amigos começaram a arreliar porque ele tinha a doença. Ficavam fazendo chacota com a cara dele. Ai ele parou mais de fazer as coisas e ficou com vergonha por causa da chacota do pessoal.

ANONYMITY

He was a happy person who lived his life as if he were healthy like other people who did not have the disease, and suddenly his friends began to pester him because of his condition. They made fun of him. So he stopped doing the things he usually did because of the mockery of his friends.

Figure 1 – Story- Drawing "Anonymity". Feira de Santana, BA, Brazil, 2018.

Financial difficulties and medication shortage discourage attendance to the Referral Center

The fact that SCD has no cure and is a chronic disease also contributes to non-adherence to treatment. In addition, financial difficulties arousing from unemployment contributed to non-adherence to treatment, as participants reported lack of transportation/money to get to the Referral Center. And the lack of medicines in the referral unit should also be mentioned. Therefore, these were the factors that contributed to discourage the users to adhere to SCD treatment, as shown in Figure 2 and the following fragments:

FALTA DE MEDICAMENTO PARA ANEMIA FALCIFORME

Então aqui é uma pessoa que tem a doença falciforme e não tem aquele interesse de fazer o tratamento devido também porque tem muitas pessoas fazem o tratamento e algumas pessoas acabam morrendo, então essa pessoa aqui já não está interessada em fazer. Devido também a falta de transporte, porque se tivesse um transporte público para levar para esse lugar seria mais fácil. E também devido à falta de dinheiro, já que não tem o transporte próprio pra isso[...]à falta de medicamento que é o Hydrea e não é fácil achar.

LACK OF MEDICATION FOR SICKLE CELL DISEASE

This is a person who has sickle cell disease and is not interested in the treatment because some people who get the treatment die. So this person is not interested in following the prescribed treatment. Another reason is the lack of transportation. If there were public transportation to the health facility it would be easier. Also, there is no money to pay for private transport [....] and the drug used in the treatment, Hydrea, cannot be easily found.

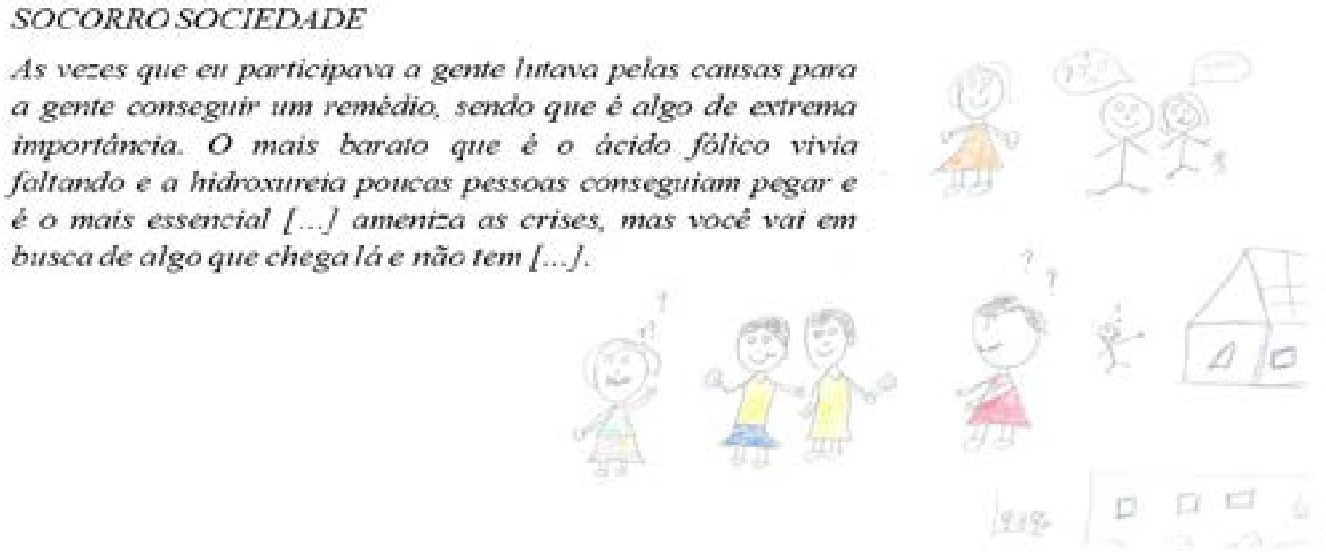
Figure 2 - Story-drawing "Lack of medications for sickle cell disease". Feira de Santana, BA, Brazil, 2018

Many people do not go to the health unit because of lack of transportation and other things. There is lack of transportation, because people cannot afford to go back and forth to the health center for treatment. (INT. 1).

[…] they also interrupt the treatment because there is always a lack of supplies or medicines. (INT. 3). There is always something missing. So they get discouraged. (INT 3)

The health unit is a space that amplifies doubts and uncertainty

According to the participants, the health unit focuses too much on the losses and limitations imposed by the disease, increasing the uncertainties about the future, and offers no guarantee that their lives will improve. See Figure 3 and the testimonies:



SOCORRO SOCIEDADE

As vezes que eu participava a gente lutava pelas causas para a gente conseguir um remédio, sendo que é algo de extrema importância. O mais barato que é o ácido fólico vivia faltando e a hidroxureia poucas pessoas conseguiam pegar e é o mais essencial [...] ameniza as crises, mas você vai em busca de algo que chega lá e não tem [...].

HELP US, PLEASE

I sometimes used to go to the health unit, and I and others fought for our right to get the medication, which is extremely important. The cheapest drug, folic acid, was always missing, and few people had access to hydroxyurea, which is the most important drug [...] and mitigates crises… so you go looking for something that is not available [...]

Figure 3 - Story-drawing “Help us, please”. Feira de Santana, BA, Brazil, 2018

[...] and others too, because it ends up frustrating our own dreams, e.g. I arrive at a place where people only talk about and report diseases and problems. And at the meetings I even heard that we must know well the people with whom we establish close relationships. I have a boyfriend, do I have to ask him if he has this disease? Can’t I have my own children? Will my children have the same problems that I have? So, I still have a lot of concerns, I don’t feel ready to look for a solution, to look for treatment. On the contrary, every time I go to the health unit I feel frustrated and more insecure than I was before the meeting. It’s something that deserves better thought and attention. (INT. 3).

Maybe it’s because I feel insecure there. I went there many times and found that it’s not just a crisis, it’s not just pain; there are other problems. At first, I was very scared and didn’t want to find out anything else. It was a little bit personal, I guess I was traumatized. (INT. 2).

Faced with so much information about possible complications, one respondent became even more afraid of SCD, ceased to go to the appointments in the health unit and withdrew from treatment, refusing to know more about the disease. So she was always worried about what might happen to her.

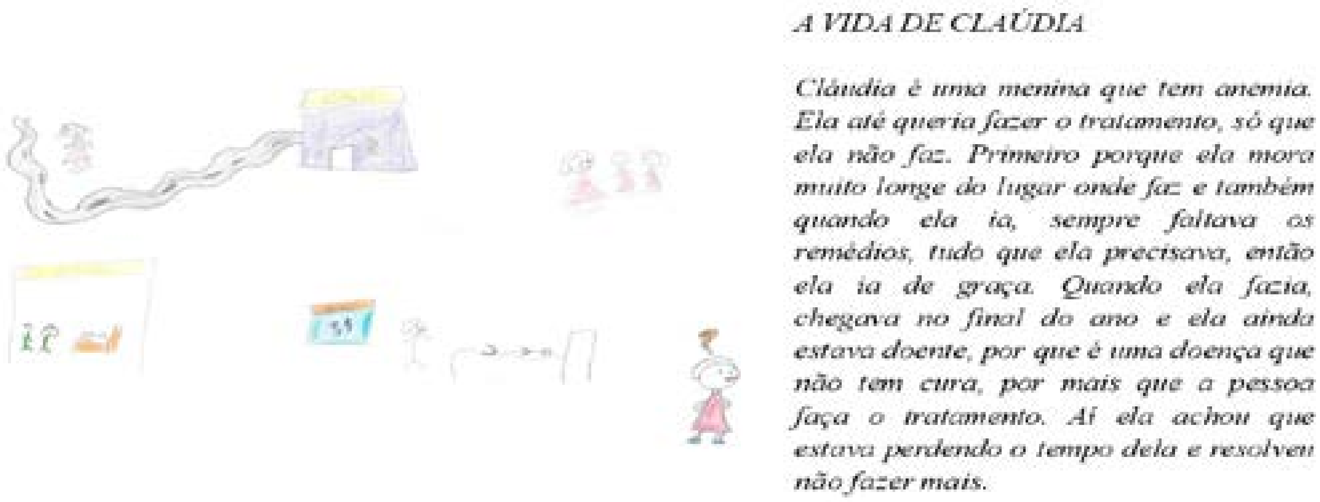
Hopelessness leads to treatment withdrawal

Obstacles encountered in the disease process, such as difficulties in accessing the health service, disorganization of the health care network, financial difficulties, and social rejection are situations that reduce the hope of the sick individuals and discourage them to seek treatment.

One of the respondents complained about the invisibility of the sickle cell disease suffering, which, for him, was evident in the ignorance of the population and lack of interest from health authorities to promote services to alleviate the adversities experienced by the sick individuals, as it can be seen below:

So, I often prefer to hide, because I don't see any improvement, I don't see any interest and I don't think it is necessary to participate in a program where there is no improvement for me. There are improvements, but the disease is there. You are still sick, you are still suffering, and the population does not realize what happened. It's about anonymity, we are hidden, camouflaged (INT 3)

In the story derived from Figure 4, the respondent highlighted the dilemmas experienced by the chronically ill, when she evaluated the fruitless efforts to manage the crises caused by the disease. This difficulty led her to question the validity of treatment adherence for her well-being and life support. It should be noted that death and fear of dying are common feelings among adults with SCD.



A VIDA DE CLAÚDIA

Cláudia é uma menina que tem anemia. Ela até queria fazer o tratamento, só que ela não faz. Primeiro porque ela mora muito longe do lugar onde faz e também quando ela ia, sempre faltava os remédios, tudo que ela precisava, então ela ia de graça. Quando ela fazia, chegava no final do ano e ela ainda estava doente, por que é uma doença que não tem cura, por mais que a pessoa faça o tratamento. Aí ela achou que estava perdendo o tempo dela e resolveu não fazer mais.

CLAUDIA'S LIFE

Claudia is a girl who has sickle cell disease. She wanted to do the treatment, but she doesn't. First, because she lives far away from the health unit and also because when she went there, the drugs she needed were missing. So it was no use going there. When she adhered to the treatment, she was still sick at the end of the year, because the disease has no cure, even for those who follow the treatment. So she thought she was wasting time and decided not to do the treatment anymore.

Figure 4 - Story-drawing "Claudia's life". Feira de Santana, BA, Brazil, 2018

DISCUSSION

The difficulties imposed by chronic illness trigger feelings of rejection and fear of the unknown, which are experienced in different ways by the individual and other members of the same family (11). The family experiences the whole process of illness along with their loved one. However, when they share the same pathology, they also experience the same repercussions and impacts on everyday life (12-13).

People with SCD must adopt strict therapeutic regimen to prevent complications. However, stigma, prejudice, discrimination, financial difficulties, lack of inputs, medicines and professionals with little knowledge about SCD in health care services favor treatment withdrawal and trigger complications (14).

In addition, the expression of the disease in the body generates shame in the sick and favors isolation and social exclusion (15), which may lead to disinterest in adherence to PD treatment. Moreover, individuals with SCD may feel ashamed and afraid of exposing their disease in health services near their home, because they may meet acquaintances in the community who may discriminate against them and exclude them from social activities. Hiding the disease can be one of the reasons for not adhering to treatment and/or giving explanations to others about care and treatment routines.

In the case of the family that participated in this study, who had a low income, made it difficult for the individuals to access the services for treatment, generating a harmful cycle for the members: when SCD is not treated, it increases fatigue and there are more painful crises and other complications that make the sick individuals less able to work. If many individuals in the same family have a debilitating chronic disease, such as SCD, this makes it more difficult for them to work and hence be capable of paying for their livelihood and going to the appointments in the health units. One study highlights that the multiple costs generated by chronic illness can exhaust the potential of family care, establishing or expanding their vulnerability if they are not assisted by health support networks (11,13).

SCD has a great impact on the lives of sick people and their families, with repercussions in various dimensions, such as social interactions, marital and family relationships, education and employment (16). Non-adherence to treatment contributes to the onset of several clinical complications and a high number of hospitalizations, causing damage and sequelae that may be irreversible to the patient. Repeated hospitalizations for SCD make patients hopeless and afraid of death and cause high costs for health services (15-17).

Low adherence to pharmacological treatment of chronic diseases has been associated with low educational level and low income, even when medications are provided at no cost to the patients by the public health system (6). In the present study, the fact that none of the family members had a job justified the non-adherence to SCD treatment, because the patients had no resources and due to the interruption of the supply of medicines in the health unit.

Adults with SCD have low self-esteem and strong feelings of hopelessness as a result of frequent pain, recurrent hospitalizations and job loss (18). These conditions make it difficult for these individuals to keep their jobs, compromising the family's livelihood.

One of the major barriers to adherence to the treatment of members of a family with SCD is that the users have questions about the therapy, which are not clarified in the health services attended. Thus, most health professionals are not adequately prepared to deliver care to people with SCD, as they lack the necessary knowledge to explain the particularities of the disease to the sick (17).

One of the participants, who was doing his/her best to forget about the SCD claimed that the health professionals kept talking about the disease and its complications and the risk of death, themes that motivated treatment withdrawal. People with SCD need to take drugs, vitamins and receive vaccines frequently in order to delay complications. However, these measures impose a strict routine of care, visits to the health service and body care that constantly remind them that they are ill (19).

The fear of dying, reported by the participants, is also highlighted in some studies with chronic patients (16). Death itself is not a problem for the patient. What scares these people is the fear of dying, originated from the feeling of hopelessness, helplessness and isolation (18).

The main issues addressed by health professionals that assist people with SCD are the negative aspects of living with the disease and its constraints. However, these issues cause the sick people to move away from these health services, in an attempt to forget that they are affected by the physical, social and emotional disabilities that this chronic illness causes. Moving forward depends on the coping with the disease, which comprises the interaction of all feelings related to expectations and support, so that suffering can be overcome (16).

Health education is an effective tool for adherence to the treatment of chronic diseases, as it allows to inform, transform and emancipate the subjects involved, contributing to good practices that facilitate health promotion and disease prevention (20). Health education, in the context of health and nursing, is made possible by the dialogue between the health professional and the patient. Thus, the individuals become aware of their health-disease situation, and can play an active role in their self-care process, contributing to adherence to SCD treatment (21).

When nurses know the reasons why people with SCD do not adhere to the treatment, they can mobilize other members of the multidisciplinary team and support network to provide emotional, instrumental and educational support, and trigger the social protection resources available to guarantee the minimum subsistence for the sick individuals (22).

One limitation of this study is the small sample size - a case study of only one family. Thus, the results cannot be generalized, nor can a cause-effect relationships be established.

CONCLUSION

The present study aimed to understand the aspects that influence non-adherence to SCD treatment in a family with several members affected by this condition. It was found that experiences of rejection, social isolation, prejudice, shame, lack of transportation to the health unit, poor financial conditions, lack of medicines/ supplies in the Referral Center, uncertainty about the treatment, lack of knowledge about the disease and its chronic character are reasons for non-adherence to the treatment of SCD.

The need to merge the approach of the disease and its complications with the approaches of care and positive experiences, encouraging hope and the construction of meanings for life in the process of chronic illness of people with sickle cell disease was emphasized in this study. The fact that five people from the same family have the disease maximizes the economic problems and impacts treatment adherence, requiring a special support from the social network and the multidisciplinary team, in order to favor treatment adherence.

This study contributes to increase the visibility of the social aspects implicated in the phenomenon of non-adherence to treatment of individuals with SCD, which may support intervention plans aimed at reducing the vulnerability of chronic patients.

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