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TEXTO & CONTEXTO ENFERMAGEM
TEXT & CONTEXT NURSING TEXTO & CONTEXTO ENFERMERÍA

KNOWLEDGE AND PRACTICE OF NURSING IN SICKLE CELL DISEASE AND HEMOGLOBINOPATHIES IN PRIMARY CARE

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ABSTRACT

Objective: to investigate the level of knowledge of nurses in Basic Health Units and their engagement in monitoring patients with sickle cell disease and other hemoglobinopathies.
Methods: this is a qualitative, descriptive-exploratory study, carried out with 12 nurses from basic health units in the municipality of Santa Luzia/Minas Gerais between August 2018 and February 2019. The semi-structured interview was the technique used for data collection, which was analyzed using Content Analysis.
Results: the analysis of the interviews emerged in the construction of three categories: understanding of sickle cell disease, risk factors, and alterations on physical examination; nursing care at the health unit according to the recommendation of the Ministry of Health; obstacles and facilitators for the tracking and identification of patients. Discourse analysis highlighted: the outstanding presence of misconceptions regarding sickle cell disease; the absence of effective follow-up of patients in the area covered by the nurse; and the non-existence of a positive sickle cell disease patient link with primary care.
Conclusion: although nursing plays a fundamental role in monitoring and assisting patients with sickle cell disease, the study revealed a significant gap between care recommendations and practice in basic health units.

DESCRIPTORS: Sickle Cell Anemia. Hemoglobinopathies. Primary health care. Nursing education. Nursing.

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CONHECIMENTO E PRÁTICA DE ENFERMAGEM NO ATENDIMENTO À DOENÇA FALCIFORME E HEMOGLOBINOPATIAS NA ATENÇÃO PRIMÁRIA

RESUMO

Objetivos: investigar o nível de conhecimento dos enfermeiros das Unidades Básicas de Saúde e o engajamento destes no acompanhamento de pacientes com doença falciforme e outras hemoglobinopatias.
Métodos: este é um estudo qualitativo descritivo-exploratório, realizado com 12 enfermeiros de unidades básicas de saúde do município de Santa Luzia/Minas Gerais entre agosto de 2018 a fevereiro de 2019. A entrevista semiestruturada foi a técnica utilizada para coleta de dados, as quais foram analisadas usando a Análise de Conteúdo.
Resultados: a análise das entrevistas emergiu na construção de três categorias: compreensão sobre a doença falciforme, fatores de risco e alterações ao exame físico; assistência do enfermeiro na unidade de saúde segundo a recomendação do Ministério da Saúde; dificultadores e facilitadores para o rastreamento e identificação dos pacientes. A análise do discurso destacou: a presença marcante de conceitos equivocados em relação à doença falciforme; a ausência de acompanhamento efetivo dos pacientes da área de abrangência do enfermeiro; e a não existência de vínculo entre paciente com doença falciforme positivo e a atenção básica ou uma lacuna significativa entre as recomendações de cuidado e a prática nas unidades básicas de saúde.
Conclusão: embora a enfermagem desempenhe um papel fundamental no monitoramento e na assistência aos pacientes com doença falciforme, o estudo revelou uma lacuna significativa entre as recomendações de cuidados e a prática nas unidades básicas de saúde.

DESCRITORES: Anemia Falciforme. Hemoglobinopatias. Atenção primária à saúde. Educação em enfermagem. Enfermagem.

CONOCIMIENTO Y PRÁCTICA DE ENFERMERÍA EN EL CUIDADO DE LA ENFERMEDAD DE CÉLULAS FALCIFORMES Y HEMOGLOBINOPATÍAS EN LA ATENCIÓN PRIMARIA

RESUMEN

Objetivo: investigar el nivel de conocimiento de los enfermeros de las Unidades Básicas de Salud y su participación en el seguimiento de pacientes con enfermedad de células falciformes y otras hemoglobinopatías.
Método: se trata de un estudio cualitativo descriptivo-exploratorio, realizado con 12 enfermeros de unidades básicas de salud de la ciudad de Santa Luzia/Minas Gerais entre agosto de 2018 y febrero de 2019. La entrevista semiestructurada fue la técnica utilizada para la recolección de datos, que fueron analizados mediante el Análisis de Contenido.
Resultados: el análisis de las entrevistas emergió en la construcción de tres categorías: comprensión sobre la enfermedad de células falciformes, factores de riesgo y alteraciones en el examen físico; atención de enfermería en la unidad de salud según recomendación del Ministerio de salud; obstáculos y facilitadores para el seguimiento e identificación de pacientes. El análisis del discurso destacó: la marcada presencia de conceptos erróneos en relación con la enfermedad de células falciformes; la falta de seguimiento efectivo de los pacientes en el área cubierta por la enfermera; y la inexistencia de un vínculo positivo entre el paciente y la enfermedad células falciformes con atención primaria.
Conclusiones: si bien la enfermería tiene un papel fundamental en la conducción del seguimiento y atención de los pacientes con enfermedad de células falciformes, el estudio reveló una brecha significativa entre las recomendaciones de atención y la práctica en las unidades básicas de salud.

DESCRIPTORES: Anemia de células falciformes. Hemoglobinopatías. Primeros auxilios. Educación en enfermería. Enfermería.

INTRODUCTION

Hemoglobinopathies are genetic diseases that affect hemoglobin, causing a structural mutation that gives rise to hemoglobin called S (Hb-S). Sickle cell disease (SCD) is the most common example of these alterations and its name refers to the shape in which the red blood cell is presented (sickle or half moon) due to the decrease in its flexibility and increase in its rigidity as a result of hypoxic conditions, where the red blood cells are located[1,2].

Present inside erythrocytes (erythrocytes), hemoglobin (Hb) is a protein that gives the red color to the blood and has as one of its main functions to transport oxygen from the lungs through the bloodstream for the normal functioning of all organs[3]. It is, therefore, an essential element in organic homeostasis and its alterations imply disorders that considerably compromise human health as it is related to chronic hemolysis and risk of vaso-occlusion[1].

This mutation is responsible for the encoding of Hb S instead of hemoglobin A (Hb A), present in adults, and occurs by modifying a nitrogenous base, adenine (A), by another, thymine (T), in the sixth codon of the beta-globin gene, causing the phenotypic change. If the mutation occurs in homozygosis, the person will receive a gene for hemoglobin S (Hb-S) from each parent. If it occurs in Heterozygosity, there will be an association of Hb S with other variants, such as SC, SD, SE diseases, or even in interaction with S/Beta thalassemia (S/B tal.) and other rarer ones[4].

If it inherits the gene for Hb S from only one parent and the gene for Hb A from the other, it will only be a carrier of the sickle cell trait (Hb-AS) and will not have the disease. However, it can transmit to the children and the family must be guided through Genetic Counseling. The term Sickle Cell Anemia is used for the most severe form and with the greatest clinical manifestation of the disease, which is Hb-SS. The mention of Sickle Cell Disease encompasses all hemoglobinopathies. Despite the particularities that distinguish them, all these combinations have similar clinical and hematological manifestations, therefore, universally, the conducts are the same for all, taking into account only the more or less severe course of each one of them[1,3,4].

It is estimated that for every thousand children born alive in Brazil, one is diagnosed with the disease, which would lead to about 3,000 cases a year. For the diagnosis of sickle cell trait, the number is even higher, with an estimate of 180,000 live births. The detection of the disease became possible after the implementation of Neonatal Screening programs aimed at early detection through the test popularly known as “Newborn Screening Test”. With the expansion of access to the screening program, there was an increase in the range of congenital diseases analyzed, defining more assertive eligibility criteria for the nationwide Neonatal Screening Program[3,5].

Due to be closer to the users’ place of residence, the Basic Health Units provide the establishment of a bond between the user and the health team, promoting a more comprehensive approach from diagnosis, assistance with a multidisciplinary team and education in health with a focus on self-care[6]. This factor is essential in reducing morbidity-mortality and increasing life expectancy for people with sickle cell disease, as they provide comprehensive care for pregnant women and newborns.

The nurse is the protagonist in the actions that begin in family planning and continue in prenatal care, delivery and puerperium[2]. In the latter, with care actions on the 5th day, the nurse supervises the nursing team in collecting material for the “Newborn Screening Test”. In addition, it performs post-heel prick counseling consultations and, if diagnosed, includes patients diagnosed with sickle cell disease and other hemoglobinopathies in the care network with a multidisciplinary and interdisciplinary team[7].

It is part of the nursing routine in the follow-up of patients with SCD and other hemoglobinopathies, screening, referral to specialized care, and after diagnosis, monitoring them for adequate use of medications, periodic examinations, and early detection of signs and symptoms of instability[8]. After screening and diagnosis, patients with SCD are followed up by a hematologist and concomitantly in primary care in the municipality.

Developing countries have a shortage in relation to the availability of specializations for nurses in the care of hemoglobinopathies[9]. Despite it, care for such patients is part of the list of nurses' activities, including within the scope of primary care.

A common failure in this chain of care is the fact that many professionals consider the treatment of such diseases to be punctual when intercurrences require hospitalization or highly complex intervention, notably in hematological centers or specialized units[10]. This perception leads to a breakdown in care, highlighting the lack of preparation and insecurity of primary care, emergency, or inpatient care professionals when these patients or family members resort to such services[11]. It is also reinforced that the link between the professional and the caregivers of patients with SCD is an essential tool to reduce the burden on the family nucleus[12].

By developing and executing various actions, primary care nurses need to know, as a priority, the profile and needs of the enrolled population. Therefore, this study aims to investigate the level of knowledge of nurses from Basic Health Units in the follow-up of patients with sickle cell anemia in their area of coverage.

METHODS

This is a descriptive observational research with a qualitative approach. The research was carried out with 12 nurses from the Basic Health Units/Family Health Program, in the municipality of Santa Luzia/Minas Gerais, which has primary care centers and approximately 221,705 inhabitants, according to an estimate by Brazilian Institute of Geography and Statistics – *IBGE*, for the year 2021[13]. Nurses of both genders listed in the network of professionals in the municipality were invited. Inclusion criteria were: active professionals with at least six months of service. The exclusion criteria were: professionals who were on vacation and therefore could not be contacted. The professionals were invited by telephone, made available by the municipal health department. The interviews were carried out until sample saturation[14]. Data collection was performed with semi-structured interviews, individually and occurred within the workplace itself, with prior scheduling and, according to the availability of the professional. The interview script consisted of the following sections: participant identification, with name (later replaced by a pseudonym), age and gender; and five semi-structured questions, described as it follows: 1) In your opinion, what would be the best definition of sickle cell disease? 2) What are the main risk factors associated with the health of patients with sickle cell anemia? 3) What alterations can be observed in the physical examination of a child with sickle cell anemia? 4) What are the actions that primary care nurses should carry out with patients with sickle cell anemia? 5) What are your difficulties regarding the care of patients with sickle cell anemia?

The interviewees' statements were recorded, with their permission, for later analysis. Respecting the ethical precepts of Resolution no. 466 of December 12, 2012, the participants signed the Informed Consent Form in two copies and, in order to guarantee confidentiality, each participant was identified with the letter E (*Enfermeira*/Nurse) followed by numerical order (E1,E2...E12). After transcription of the data, the recordings were erased to maintain the secrecy and integrity of the interviewees.

Data collection took place from August 2018 to February 2019. To determine the reach of theoretical saturation in the primary sources, five procedural steps were followed: Record of raw data (primary sources) – in total, 12 interviews. From the beginning of data collection, the interviews were recorded with the permission of the interviewees and immediately transcribed in full; Immersion in the data: a floating reading of the data obtained through the interviews was performed as they were carried out; Compilation of individual analyzes of each interview and thematic grouping; Allocation of themes and types of statements in a table: the presentation of data in a table allowed the identification of the regularity of the findings in the testimonies, according to the themes, and the verification of the consistency of the statements; Verification of the theoretical saturation of the data by identifying the absence of new elements in each grouping[14–15]. The analysis of the collected material followed a rigorous process in view of the phases defined by Bardin, such as: Organization; Pre-analysis; Exploration of the material, and Treatment of the results[16].

RESULTS

The final sample consisted of 12 participants, all female. The graduation time ranged from one year and three months to eleven years, with 6 interviewees (50%) having between 1 year and 3 months and 6 years of training. From these ones, only 2 had specialization. The other 6 interviewees (50%) had between 7 and 11 years of training, and only 2 had no specialization. Among the participants with a postgraduate degree, none had a specialization related to the field of hematology.

The time working in primary care ranged from one (1) to nine (9) years, with 7 respondents (58%) having between 1 and 4 years and 5 (42%) having between 5 and 9 years of experience. Regarding the time of occupation of the current position of nurse in the basic health unit, 4 interviewees (33%) had 1 to 3 years; 5 (42%) were 4 to 5 years old; and 3 (25%) were 6 to 9 years old.

The analysis of the interviews resulted in the construction of three categories: understanding about sickle cell disease, risk factors and changes in the physical examination; nursing care at the health unit according to the recommendation of the Ministry of Health; obstacles and facilitators for the tracking and identification of patients.

Understanding about sickle cell disease, risk factors and alterations to the physical examination

Regarding to understand the disease, some professionals demonstrated difficulties in mastering the topic. For example, there were speeches in which the symptoms mentioned were confused and associated with the diagnosis of other diseases such as the immune system or coagulopathies.

It is a disease in the case of the immunobiological system, an alteration of the red blood cell, which, instead of being C-shaped, is usually S-shaped (E8).

In fact, we still know little, right? Because it is a disease, I believe, I know it is an autoimmune disease. A genetic disease passes from parent to child. That is what I know (E11).

What I do know. It is a problem with red blood cells that have a sickle shape. I think she has platelet difficulties, clotting problems, things like that, I do not know (E3).

[...] *that is why they say sickle shape, like this: sickle cuts, right, and it hurts a lot* (E10).

I know what I have studied in college, and I have been a nurse in the unit for eight years and I've never participated in any specific training (E4).

Regarding risk factors and alterations on physical examination, the majority showed some knowledge, however, as previously mentioned, there were still associations with other diseases. It is worth mentioning that the professionals' lack of knowledge about the warning signs can compromise

the necessary assistance in a timely manner to these patients. To be aware of this seriousness, an episode of fever must be interpreted as a dangerous situation and its diagnosis must be carried out to allow immediate therapy.

It is a patient who may have a problem, I think it is chest syndrome, he has difficulty breathing, a risk for stroke, splenic sequestration, and there is a risk for vision. They have a lot of joint pain, due to the low oxygenation due to this hemoglobin format that ends up making it difficult to transport oxygen and in this case, it is only with morphine (E5).

Related to blood itself, clotting, and anemia perhaps. I figure out there is more risk of bleeding (E3).

Risk factors are closely linked to his treatment. If he is assiduous, it has a lot to do with the production of red cells. Risk factors are closely linked with the patient, with the patient's immunity in the production of cells [...]. *It will depend a lot on the evolution of the disease, the patient's time and adequacy to the treatment* (E7).

As I spoke to you. My knowledge of this patient is limited. A patient like the one who arrives for us, we receive the diagnosis and he/she is referred to the municipality, for the Nupad program (E9).

In my unit, they do not consult. My bond with the Sickle Cell Disease staff is due to the active search when they miss the Nupad consultation, which is a monthly control (E4).

The inconsistencies and uncertainties identified in these testimonies reveal a lack of preparation and lack of understanding on the part of professionals in the early identification of signs and symptoms of aggravation in patients with sickle cell disease and hemoglobinopathies.

Nurse assistance at the basic health unit according to the Ministry of Health

The second category refers to the assistance provided by nurses at the health unit, as recommended by the Ministry of Health. On the subject, some demonstrated lack of knowledge, even considering that it is performed only by the Blood Center.

It is difficult to pick up a patient with sickle cell anemia since the heel prick test, which is the screening test on the fifth day, when it is positive, it is already referred to Hemominas and that is where he will receive follow-up and treatment (E8).

I don't know. For us here, when the child arrives, it is more or less a flow, a protocol, you know. So when it arrives and this exam comes with alteration, it comes from Nupad, we refer it to the specialist, then he/she does the flow there, with us in the unit, the patient is not followed up (E9).

From sickle cell anemia? There is no monitoring in the unit. The follow-up is with the specialist, right, and the hematologist. Then they don't come, like I said, there's no follow-up here at the health center, you know (E12).

Here we have a pediatrician, who monitors normal child care, and patients follow up on sickle cell anemia at Hemominas (blood center) (E2).

It has already been demonstrated that there is a barrier that prevents people with sickle cell disease from seeking the primary care health service, moving directly to secondary care, represented by the blood center.

Regarding consultations, they report that they are carried out according to the routine schedule recommended for all children, with no follow-up of the recommended interval in a specific calendar for children with sickle cell disease. As it is a disease that confers immunological vulnerability, the vaccination schedule must go beyond the basic schedule vaccines (BCG, Hepatitis B, Polio, Rotavirus, Pentavalent, Pneumo 10, Meningitis C, Yellow Fever, Triviral, Chickenpox and Hepatitis A), special ones like Pneumo 23 (from 2 years) and Influenza (from 6 months)[17]. The professionals showed lack

of knowledge about the use of medications used by these patients, which implies a lack of monitoring of correct use. Regarding the schedule of special vaccines, it was limited to the basic schedule recommended for all children, not following the Ministry of Health recommendations for these patients.

Difficulties and facilitators for the tracking and identification of patients

As for the tracking and identification of patients in the area covered by the units, some demonstrated that they were aware of the existence of these patients. Others categorically stated that it did not exist, while others remained in doubt.

Yes. I have 2 cases (E11).

Yes, there are patients here in the sickle cell anemia region (E8).

I know only one (E1).

No. Not by heart (E12).

Not from anemia (E9).

In other units I had already had it, but here that I remember, I don't have it (E2).

Not that I know (E4*).*

No. As far as I know, there is none (E3).

Regarding the factors that hinder and/or facilitate this follow-up, most responses referred to the difficulty that patients have in relation to adherence and adequacy to treatment. For them, the fact that patients do not commit, it makes the work of the unit's professionals more difficult.

What makes it difficult is adherence. They don't go to the unit because they know that there isn't a hematologist, there's only a clinician. It's easier for us to look for them, do an active search when Hemominas asks (E8).

The first thing that I find difficult is when the patient does not adapt to his/her treatment. Often they do not accept the pathology and this makes it difficult because you have to try to show that they need help (E7).

People who doesn't want to take care of themselves properly, because there are patients that they don't give themselves to, they don't adapt (E6).

Others still mentioned the lack of knowledge about the disease as a hindrance.

What really makes it difficult is the lack of knowledge about the disease. We delve a lot into what we have, what we follow, what we see. What we don't see ends up being neutral. It's a lot, it's a very wide range of diseases that we end up having to monitor (E9).

It is a somewhat small population, and many times, we are more concerned about infectious diseases than these hematopathological diseases (E8).

It was also mentioned by some that the lack of communication between health services compromises this patient care.

I think that the reference service fails because there is no exchange of knowledge with the unit. Maybe if they sent us a copy of the chart. We don't have this exchange of information, so the post ends up staying, just to give the diagnosis when it's a child (E4).

I think that having a counter-reference from the hospital to primary care would help in this care. There is always this lack of communication from one attention to another, which makes it very difficult (E3).

Other obstacles were mentioned, such as the absence of a physical address (E11), the lack of professional training (E5) and the fact that users do not seek the unit, since for them; the follow-up is carried out at the Blood Center (E12).

The facilitators referred to the ease of access of users to the units.

What facilitates is the easy access to our unit (E6).

I think that access is not difficult in the sense that, every time something appears, we mobilize, and get in touch; it is not difficult to get in touch with the municipal health secretary (E5).

Others indicated the possibility of creating groups with patients for conversations and exchange of experiences (E8), acceptance of patients in relation to the pathology (E7), identification of patients in the coverage area (E3), and early diagnosis through screening neonatal (E4).

DISCUSSION

Notwithstanding the recommendations of the National Policy for Comprehensive Care for People with Sickle Cell Disease, the data from this study showed a gap between the recommendation and the evidenced reality. Permanent education should be considered one of the focuses for training all professionals involved. When low levels of knowledge are evidenced, we understand that this action is not being carried out or its implementation is precarious. The deficits of knowledge evidenced by the study may reveal an academic gap and also a continuing education gap in the qualification chain of health professionals[18]. In addition, it is a challenge to act in primary care, since it has a significant variety of comorbidities and assigns professionals to monitor and care for these patients. In the case of care for sickle cell anemia, the interviewed nurses did not have specific training and this factor is possibly involved in the gaps of observed knowledge[19].

Unknown or even neglected by many professionals, SCD interferes in various aspects such as social interaction, marital and family relationships, education, employment, among others[1,11]. Originating from the African continent, sickle cell disease is one of the most common autosomal recessive hereditary diseases in Brazil and is considered a public health problem. Its name refers to the format in which the red blood cell is presented (sickle or half moon) due to the decrease in its flexibility and increase in its rigidity because of the hypoxic conditions in which they are found[1,3,20]. Despite this significant impact and the fact that our country has a care program for this pathology, the testimonies of the interviewed nurses highlighted an incorrect interpretation of the pathological process and even of their performance in the health care of this patient. It is important to discuss this situation within a health context in which the participants' primary care services make part. Such difficulties may be associated with the organizational level of health services, the scarce experience in care practice for this pathology and the lack of support from a multidisciplinary team[21]. The lack of adequate knowledge about SCD care by nurses has already been observed in other countries[22–23].

The early detection of the disease through neonatal screening aims to reduce injuries, increase life expectancy and improve its quality and its follow-up must be carried out by a multidisciplinary team and permeate the entire Health Care Network, which includes Primary and Specialized Care, Urgency and Emergency and the Hospital[3,8]. Therefore, it is necessary for nurses to have knowledge, skills and competences to recognize warning signs and symptoms in patients with SCD[11]. However, most ones demonstrated mistaken knowledge when they mentioned associations with other diseases. It is worth mentioning that the professionals' lack of knowledge about the warning signs can compromise the necessary assistance in a timely manner to these patients. To be aware of this seriousness, an episode of fever must be seen as a dangerous situation, the diagnosis must be carried out and deepened as soon as possible and therapy must be started immediately[24–25].

According to the Ministry of Health, sickle cell disease has clinical manifestations that compromise the daily lives of its patients. Pain crises are the most frequent and commonly the first clinical manifestation. We also mention dactalitis (hand-foot syndrome), ocular complications, anemia, acute chest syndrome, jaundice, splenic sequestration crises, stroke, leg ulcers and osteonecrosis, priapism, biliary lithiasis, recurrent infections among others[5,26]. Despite the particularities that distinguish the association of Hb-S with other variants, all these combinations have similar clinical and hematological

manifestations, therefore, universally, the conducts are the same for all, taking into account only the most severe course of each one[5,27].

The entire Health Care network must carry out the follow-up of patients with SCD. Basic Health Unit nurses play an extremely important role in conducting this follow-up[25] since they are present from the child's mother's prenatal care, childcare and receiving the diagnosis, about possible complications that can be identified and, if necessary, forwarded to the reference service[11,25].

As exemplified by one of the interviewees (E8) in our study, there is little representation of this population group in health networks, which raises an important point to be rethought – the inclusion of patients and family members who deal with PA in their daily lives. A barrier has already been described which makes the person with sickle cell disease not seek primary care health service, moving directly to secondary care, represented by the blood center[28]. They also reported that the consultations for growth and development, the special vaccine schedule, monitoring, and knowledge about the use of specific medications should be priorities for nurses to achieve quality care in this area.

Regarding consultations, the interviewees reported that they are carried out according to the routine schedule advocated for all children, with no follow-up of the recommended interval in a specific calendar for children with SCD. The professionals showed lack of knowledge about the use of medications used by these patients, which implies a lack of monitoring of correct use. In relation to the schedule of special vaccines, it was limited to the basic schedule recommended for all children, not following the recommendations of the World Health Organization for these patients[8,27].

Active in permanent education, the nurse must guide patients on the practice of self-care, care for the environment and other triggering factors of acute episodes. This activity is essential for them to avoid complications and achieve quality of life[11,25]. In the basic health unit, the control consultations must follow a periodicity, except for the need for extraordinary consultations, being established up to one year: monthly appointments, from one to five years old: appointments every three months and from five years old: appointments every six months. In addition to providing all the services related to the follow-up of these patients, the Basic Health Unit also needs to form and strengthen the bond between patients, their families and the team. This factor is essential to facilitate understanding of the disease, anticipate risk situations and avoid complications that require hospital admission[9,27,29].

The performance of primary care in monitoring these patients is still very concentrated in blood centers, but this service does not replace the role of primary care. This perception about the treatment of the disease is traditionally seen as being the responsibility of the hematological centers by themselves[11], and the intermediate levels of health care are unaware of or even ignore the disease within the line of care. In this way, the breakdown of care can occur when patients or family members resort to basic care services, urgency or need care in a hospitalization unit[11]. On this occasion, there is a greater risk of insecurity and poor quality of care.

Among the attributions of primary care are performing the heel prick test on 100% of newborns in its area, the mapping of people with sickle cell disease in the coverage area, the promotion and participation of health education actions for self-care, the inclusion of people with sickle cell disease in the various programs developed at the Basic Health Unit, to keep up-to-date knowledge about the pathophysiology of sickle cell disease, clinical aspects, as well as the socioeconomic and cultural characterization of the affected population. The nursing profession has a unique role in this process, through the mobilization of health agents, families, and patients in the community in order to monitor effectively the public of patients with SCD and hemoglobinopathies.

This study has gaps and sample limitations, as it was carried out in a single municipality and with a restricted sample number. These factors can influence the generalization of the findings of this research.

CONCLUSION

This research revealed the presence of a knowledge deficit among the nurses interviewed in relation to sickle cell disease, both in the knowledge of the biological processes of the disease and in the application of segment protocols. Such findings allowed understanding the relevance of knowledge about the disease, as well as the importance of conducting its follow-up. Nurses in Basic Health Units play a fundamental role in this management, as the bond created between these professionals and patients/family members/community provides an effective approach in their daily work, which should facilitate health education and the promotion of self-care.

It is important to highlight the need for higher education training institutions to include disciplines, training and updating courses in the area of assistance to hemoglobinopathies in their curricular components. In addition, the health network is co-responsible for the quality of care for these patients and, therefore, it needs to promote changes in care flows and train professionals involved in both primary and specialized care. Using innovative strategies to prepare professionals can be an assertive way to be used by both educational institutions and public health management services[30].

Gaps in knowledge and slackening of follow-up only increase the distances for the prevention of injuries and quality of life in patients with SCD. The nurse requires a more critical look, beyond inherent skills of the profession itself, to provide good health care. Although, the perception of gaps in knowledge and care practice reveals that there is still much to be done, and investments in training in this area are necessary and well reasoned. It is also suggested strategies for a systematic follow-up of such patients, avoiding the absence of data about their health in primary care.

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