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Sickle cell disease: caring for people with leg ulcers in heal-thcare services

Doença falciforme: cuidado com pessoas com úlcera da perna nos serviços de atenção à saúde

Enfermedad falciforme: cuidado de personas con úlcera de pierna en los servicios de atención a la salud

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ABSTRACT

Objective: to identify the location and direct care received by people with leg ulcers due to sickle cell disease in health care services. **Method:** a cross-sectional study carried out in 11 centers from August 2019 to April 2020. The study included 72 people with active leg ulcers. The study was approved by the Research Ethics Committee. **Results:** a total of 91.7% of the participants had sickle cell anemia, with a median of three years of ulcer existence; 77.8% were recurrent; 40.3% bought the supplies; 66.7% changed their own dressings at home; 52.8% did one or more changes a day; 45.8% of the treatments were prescribed by physician; 37.5% were ointments (collagenase or antibiotics); and 89% did not use compression to manage edema. **Conclusion**: most of the participants were not included in the Health Care Network for ulcer treatment and did not receive systematized care or appropriate supplies.

Descriptors: Nursing Care; Enterostomal Therapy; Leg Ulcer; Anemia, Sickle Cell; Health Care Levels.

RESUMO

Objetivo: identificar o local e os cuidados diretos recebidos por pessoas com úlceras da perna por doença falciforme nos serviços de atenção à saúde. **Método:** estudo transversal, realizado em 11 centros, no período de agosto de 2019 a abril de 2020. Fizeram parte do estudo 72 pessoas com úlcera da perna ativa. O estudo foi aprovado pelo Comitê de Ética em Pesquisa. **Resultado:** apresentavam anemia falciforme 91,7% dos participantes, com mediana de três anos de existência da úlcera; 77,8% eram redicivantes; 40,3% compravam os insumos; 66,7% trocavam o próprio curativo no domicílio; 52,8% realizavam uma ou mais trocas diárias; 45,8% dos tratamentos foram prescritos pelo médico; 37,5% eram pomada (colagenase ou antibiótico); 89% não utilizavam compressão para o manejo do edema. **Conclusão**: a maioria dos participantes não estava inserida na Rede de Atenção à Saúde para o tratamento da úlcera, e não recebia assistência sistematizada e nem insumos apropriados.

Descritores: Cuidados de Enfermagem; Estomaterapia; Úlcera da Perna; Anemia Falciforme; Níveis de Atenção à Saúde.

RESUMEN

Objetivo: identificar el lugar y los cuidados directos recibidos por personas con úlceras de pierna por enfermedad falciforme en los servicios de atención a la salud. **Método:** estudio transversal, realizado en 11 centros, en el período de agosto de 2019 a abril de 2020. Participaron 72 personas con úlcera de pierna activa. El estudio fue aprobado por el Comité de Ética en Investigación. **Resultado:** presentaban anemia falciforme 91,7% de los participantes, con una mediana de tres años de existencia de la úlcera; 77,8% eran recidivantes; 40,3% compraban los insumos; 66,7% cambiaban su propio vendaje en el domicilio; 52,8% realizaban uno o más cambios diarios; 45,8% de los tratamientos fueron prescritos por el médico; 37,5% eran pomada (colagenasa o antibiótico); y 89% no utilizaban compresión para el manejo del edema. **Conclusión:** la mayoría de los participantes no estaba integrada en la Red de Atención a la Salud para el tratamiento de la úlcera, y no recibía asistencia sistematizada ni insumos apropiados.

Descriptores: Cuidados de Enfermería; Estomaterapia; Úlcera de la Pierna; Anemia de Células Falciformes; Niveles de Atención de Salud.

INTRODUCTION

Sickle cell disease encompasses a group of hemoglobinopathies characterized by the presence of mutations or deletions in the β-globin gene, which leads to the production of hemoglobin S. In this condition, red blood cells undergo sickling and hemolysis when deoxygenated, resulting in vaso-occlusion and associated ischemia. In addition, this disease is characterized by recurrent episodes of intense acute pain and other complications, such as leg ulcers[1].

Ulcers occur exclusively on the legs and can be single or multiple. Characterized by recurrence and a prolonged healing process, they induce significant chronic pain, contributing to depression, disability and unemployment[2].

Over time, the understanding of the pathophysiological processes underlying the development of leg ulcers in individuals with sickle cell disease has evolved considerably. Currently, a range of theories have been proposed to explain this complex phenomenon, including vaso-occlusion, hemolysis, venous incompetence, hypercoagulability and thrombosis, autonomic dysfunction and genetic elements. Each of these proposals contributes to understanding the

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intricate interplay of factors that can precipitate and sustain ulcer development, as well as the correct choice of treatment[3].

Providing specialized care for people with sickle cell disease, especially those with ulcers, becomes a challenge, considering the population with this condition is affected by other comorbidities. These people often have complex medical issues. Allied to this situation is the lack of an organized healthcare network and specialists to provide assertive care.

In this context, although there is no standardized protocol for the treatment of these ulcers, it is essential to address both the topical treatment and the underlying pathophysiology of the condition in order to improve the quality of life and prognosis of patients and achieve the outcome, which is healing[4]. Another fact that reinforces the need to identify how these people are being cared for is the high prevalence of ulcers in individuals living in various regions of the world. In countries such as Ghana, Italy and the United States, the prevalence reaches 10.8%[5]; in Saudi Arabia, it is 8%[6]; while in Brazil it reaches 1.4%[7].

The lack of knowledge about how patients with leg ulcers are treated in health services is an important gap that this study aims to fill. Knowing the specific barriers that patients face in accessing health services and the appropriate supplies for treating ulcers can provide valuable ideas for improving care. Identifying the direct care these patients receive is crucial to evaluating the effectiveness of treatment protocols, as well as determining areas for improvement. This understanding can lead to more targeted, personalized and efficient interventions to improve the quality of life of these patients.

This study aimed not only to fill a knowledge gap, but also to contribute to the optimization of healthcare offered to patients with sickle cell disease who have leg ulcers. By identifying unmet needs and existing barriers, it is hoped that the study's findings can support health policies, clinical guidelines and intervention strategies that will improve the lives of these patients and contribute to more effective and comprehensive treatment.

Therefore, the main objective of this study was to identify the location and direct care received by people with leg ulcers due to sickle cell disease in health-care services.

METHOD

This is a descriptive observational study based on the guidelines of the tool Reporting of Observational Studies in Epidemiology (STROBE)[8]. The research was conducted at a reference foundation in hematology and hemotherapy, made up of 11 units located in different cities in Minas Gerais, which offer outpatient care to individuals with sickle cell disease. In that state, all patients with this disease are registered for care at this foundation.

The inclusion criteria for the study were participants of both sexes, with a diagnosis of sickle cell disease, aged 18 or over, up-to-date registration with the Foundation, presence of an active leg ulcer, with a chronic ulcer. All patients with ulcers of another etiology on the leg or in another region of the body were excluded from the study.

Data was collected between August 2019 and April 2020. Due to the limitations of the electronic medical records system, it was not possible to identify patients with leg ulcers through this system. In response to this limitation, professionals from different areas in the institution, such as physicians, nurses, psychologists and social workers, with the associations of people with sickle cell disease in Minas Gerais, played a crucial role in identifying these individuals.

For eligibility, a survey was carried out among the 5,379 people over the age of 18 registered at the 11 units to identify all those who had active leg ulcers. This strategy is called a census.

When people with ulcers were identified during their appointments with the hematologist, social worker or nurse, the researchers took the opportunity to collect the data. As a result, 77 individuals with active leg ulcers were identified, 72 of whom agreed to take part in the study.

Data was collected through structured face-to-face interviews conducted by researchers and/or nurses at the outpatient clinic, in addition to the leg assessment. A form was used to guide the researcher in conducting the interview and assessing the patients. The questions covered the variables relevant to the investigation, including clinical aspects such as smoking, alcoholism and the subtype of sickle cell disease. The level of pain in the ulcer was assessed using the Numerical Rating Scale and categorized as "no pain" (zero), "mild pain" (≤ 3), "moderate pain" (> 3 ≤ 6) and "severe pain" (> 6 ≤ 10)[9].

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Other aspects were also analyzed, such as the presence of edema in the lower limbs, a previous history of ulcers, the age at which the first ulcer appeared, the number of active ulcers, the length of time the ulcer had existed in years and recurrence. The presence of edema was positive if the depression (sign of Godet) was formed by compression of the fingers. If there was more than one ulcer, the oldest of the current ones was considered.

With regard to the topical treatment of the ulcer, the variables were the product used; the person responsible for indicating the treatment; the location and frequency of dressing changes; the supply of the material and the therapy for managing leg edema. In addition, the study explored variables related to the Health Care Network and the support network, including the presence of supplementary health insurance, periodic follow-up with a hematologist and with the Family Health Team, as well as periodic visits by the Community Health Worker.

The statistical analysis was carried out using the Statistical Package for the Social Sciences (IBM SPSS®), version 19. software (Chicago, Illinois, United States). The study variables were subjected to descriptive statistical analysis, using central tendency and dispersion measures. The representation included the percentage, which expressed the relative frequency of occurrences, the mean accompanied by the minimum and maximum values and the median together with the quartiles.

The research protocol for this study received ethical approval in 2019 from two Research Ethics Committees: that of the proposing institution and that of the co-participating institution, where the study was conducted. All study participants signed an informed consent form.

RESULTS

The study included 72 people with active leg ulcers, with the clinical variables shown in Table 1.

Table 1: People with sickle cell disease and leg ulcers, by gender, according to clinical variables (n=72). Minas Gerais, MG, Brazil, 2020.

Variables	Male n (%)	Female n (%)	Total n (%)
Subtype of sickle cell disease			
HbSS	33 (45.8)	33 (45.8)	66 (91.7)
Hb SbetaTALA	1 (1.4)	1 (1.4)	2 (2.8)
HbSSC	-	2 (2.8)	2 (2.8)
Did not know	1 (1.4)	1 (1.4)	2 (2.8)
Categorized pain score*			
No pain	7 (9.7)	6 (8.3)	13 (18.1)
Low (≥ 1 to ≤ 3)	5 (6.9)	4 (5.6)	9 (12.5)
Moderate (≥ 4 to ≤ 6)	12 (16.7)	9 (12.5)	21 (29.2)
Intense (≥ 7 to ≤ 10)	11 (15.3)	18 (25.0)	29 (40.3)
Age of first ulcer, years+			
≤ 10	1 (1.4)	3 (4.2)	4 (5.6)
> 10 to ≤ 20	21 (29.6)	20 (28.2)	41 (56.9)
> 20 to ≤ 50	12 (16.9)	14 (19.7)	26 (36.6)
Previous history of ulcer			
No	1 (1.4)	3 (4.2)	4 (5.6)
Yes	34 (47.2)	34 (47.2)	68 (94.4)
Number of active ulcers			
1	20 (27.8)	23 (31.9)	43 (59.7)
2	9 (12.5)	10 (13.9)	19 (26.4)
3-10	6 (8.4)	4 (5.6)	10 (13. 9)
Length of time the ulcer has existed, in years			
≤ 0.5	10 (13.9)	7 (9.7)	17 (23.6)
>0.5 to ≤2	9 (12.5)	8 (11.1)	17 (23.6)
>2 to ≤ 5	6 (8.3)	8 (11.1)	14 (19.4)
>5 to ≤ 10	6 (8.3)	5 (6.9)	11 (15.3)
>10 to ≤ 45	4 (5.6)	9 (12.5)	13 (18.1)
Recurrence			
No	7 (9.7)	9 (12.5)	16 (22.2)
Yes	28 (38.9)	28 (38.9)	56 (77.8)
Total	35 (48.6)	37 (51.4)	72 (100)

Caption:* The highest pain score was considered when more than one ulcer was present; +the variation in the total of n is due to missing data.
HbSS: sickle cell anemia; Hb SbetaTALA: hemoglobin S beta-thalassemia; HbSSC: hemoglobin S SC.

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Sickle cell disease of the sickle cell type (HbSS) was present in 66 (91.7%) people, and 68 (94.4%) had an ulcer in the past which had already healed. The median age of the first ulcer was 18 years (quartile 1 = 15; quartile 3 = 27), and 41 (56.9%) had their first ulcer between the ages of 10 and 20, 56 (77.8%) had recurring ulcers and 43 (59.7%) had only one active ulcer.

The median time the ulcer had existed was three years (quartile 1 = 0.53; quartile 3 = 7.7), with 17 (23.6%) having existed for six months or less. With regard to pain, the median pain score was 3 (quartile 1 = 0.53; quartile 3 = 7.75); 29 (40.3%) reported severe pain.

Regarding the use of secondary and tertiary care, 62 (86.1%) underwent regular follow-up with a hematologist at blood centers, and 22 (30.6%) had been hospitalized at least once in the last 6 months. With regard to Primary Health Care, 30 (41.7%) reported that they were regularly followed up by a Family Health Team, and 37 (51.4%) received at least one monthly visit from a Community Health Worker.

With regard to direct care for ulcers, 48 individuals (66.7%) were treated exclusively at home, and 11 (15.3%) combined home care with care at Primary Health Care Center. In addition, nine participants (12.5%) received treatment in outpatient secondary care services, of which 6 (8.3%) came from public institutions and 3 (4.2%) from private ones. Four people (5.6%) received their dressings exclusively at Primary Health Care Center.

Table 2 shows the products used in the topical treatment of ulcers.

Table 2: Topical ulcer treatment and those responsible for the indication (n=72). Minas Gerais, Brazil, 2020.

	Indication of treatment				
Topical treatment	**Nurse n (%)**	**Nursing technician n (%)**	**Physician n (%)**	**Others* n (%)**	**Total n (%)**
Exposed ulcer	-	-	1 (1.4)	1 (1.4)	2 (2.8)
EFA	2 (2.8)	-	3 (4.2)	2 (2.8)	7 (9.7)
Collagenase	4 (5.6)	-	11 (15.3)	1 (1.4)	16 (22.2)
Hydrogel	-	1 (1.4)	5 (6.9)	-	6 (8.3)
Interactive coverings†	11 (15.3)	-	1 (1.4)	2 (2.8)	14 (19.4)
Popular belief	-	-	-	1 (1.4)	1 (1.4)
Saline solution and gauze‡	1 (1.4)	-	2 (2.8)	1 (1.4)	4 (5.6)
Herbal ointment	-	-	1 (1.4)	3 (4.2)	4 (5.6)
Topical corticosteroid	-	-	-	1 (1.4)	1 (1.4)
Antifungal	-	-	-	1 (1.4)	1 (1.4)
Topical antibiotic	1 (1.4)	-	8 (11.1)	2 (2.8)	11 (15.3)
Moisturizing cream	1 (1.4)	-	1 (1.4)	1 (1.4)	3 (4.2)
Unna§ bandage	2 (2.8)	-	-	-	2 (2.8)
Total	22 (30.6)	1 (1.4)	33 (45.8)	16 (22.2)	72 (100.0)

Notes: *Patient themselves, family or friends; † hydrocolloid, alginate, foam and charcoal; ‡ gauze moistened in physiological solution; § part of the Unna bandage used as primary cover.
EFA: essential fatty acids.

In the group of products used for topical ulcer treatment, 16 people mentioned collagenase (22.2%), 14 interactive dressings (19.4%) and 11 antibiotic ointments (15.3%). These products were recommended by professionals and other individuals.

The interval at which participants changed their dressings varied from once a week to more than twice a day, depending on the treatment used (Table 3).

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Table 3: Topical ulcer treatment and number of changes (n=72). Minas Gerais, Brazil, 2020.

Topical treatment	Frequency of dressing changes					
	Not applicable n (%)	≥ 2 times/day n (%)	1 time/day n (%)	2 times/week n (%)	1 time/week n (%)	Total n (%)
Exposed ulcer	2 (2.8)	-	0	-	-	2 (2.8)
EFA	-	3 (4.2)	4 (5.6)	-	-	7 (9.7)
Collagenase	-	5 (6.9)	11 (15.3)	-	-	16 (22.2)
Hydrogel	-	-	5 (6.9)	1 (1.4)	-	6 (8.3)
Interactive coverings	-	-	0	9 (12.5)	5 (6.9)	14 (19.4)
Popular belief	-	1 (1.4)	0	-	-	1 (1.4)
Physiological solution*	-	1 (1.4)	3 (4.2)	-	-	4 (5.6)
Herbal ointment	-	-	4 (5.6)	-	-	4 (5.6)
Topical corticosteroid	-	-	1 (1.4)	-	-	1 (1.4)
Antifungal	-	-	1 (1.4)	-	-	1 (1.4)
Topical antibiotic	-	3 (4.2)	7 (9.7)	1 (1.4)	-	11 (15.3)
Moisturizing cream	-	1 (1.4)	2 (2.8)	-	-	3 (4.2)
Unna† bandage	-	-	-	1 (1.4)	1 (1.4)	2 (2.8)
Total	2 (2.8)	14 (19.4)	38 (52.8)	12 (16.7)	6 (8.3)	72 (100)

Notes: *Gauze moistened in physiological solution; † part of the Unna bandage used as a primary covering.
EFA: essential fatty acids.

For the presence of edema, 55 (76.4%) people had edema in at least one of their lower limbs, 49 (89%) did not use edema control therapy, 2 (3.7%) used compression stockings and 4 (7.3%) Unna boots. With regard to supplies for ulcer treatment, 29 (40.3%) people bought all the necessary material, 19 (26.4%) received it exclusively from the public network, 18 (25%) received part of it from the public network and had to buy part, 2 (2.8%) had all the material guaranteed by the private network, and 2 (2.8%) depended on donations from third parties.

DISCUSSION

The persistence of ulcers and their recurrence in people with sickle cell disease is a reality that causes disabling and long-lasting pain, resulting in consequences such as depression, disability and unemployment. Although a considerable proportion of these patients with ulcers and recurrence achieve healing, it is important to note that many live with these wounds for more than two decades, and some do not achieve healing. Considering the possibility of amputation, in certain cases, is considered in order to improve the quality of life of these patients[2].

The diversity of sickle cell disease subtypes, resulting from the inheritance of mutant hemoglobin S (HbS), can manifest in the homozygous state, known as sickle cell anemia (HbSS), or in combination with other abnormal hemoglobins, giving rise to compound heterozygotes, such as SC (HbSSC), S beta-thalassemia (HbS betaTALA), S alpha-thalassemia (HbSS alphaTALA), SD (HbSSD) and SE (HbSSE). Individuals with sickle cell anemia have a greater clinical impact, including a higher incidence of leg ulcers[10]. This highlights the importance of considering genetic heterogeneity when planning interventions and treatment strategies.

Although leg ulcers are more prevalent in men, reaching a ratio of 2:1 in certain studies[5,6,11], there was a higher occurrence of leg ulcers in women in the study carried out. This reversal of the usual trend highlights a particularity that is relevant to understanding the factors associated with this manifestation in female patients.

Most ulcers initially appear in people between the ages of ten and 20[5], highlighting the early impact of the disease, especially during school age. This precocity can have possible socio-economic implications for the schooling and income of these patients. In addition, individuals with a previous history of ulcers had a higher incidence of ulcers (11.9%) than those with no history (0.52%)[10].

Another distinct phenomenon in ulcers caused by sickle cell disease is pain, characterized by an intensity reported by patients as disabling, continuous and prolonged, different from vaso-occlusive pain.[12] This differentiation of vaso-occlusive pain from the disease highlights the unique nature of the pain associated with these ulcers, reinforcing the need for specific therapeutic approaches for the effective management of this symptomatology.

The Brazilian Ministry of Health document "Sickle Cell Disease: Basic Guidelines for the Line of Care"[13] recommends that Primary Health Care Centers are integrated with other levels of care, providing comprehensive care for these people. The results of this study confirmed that the majority of people with leg ulcers due to sickle cell

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disease attended scheduled hematology appointments at the Hemominas Foundation. However, less than half reported being followed up periodically by the Family Health Teams.

This finding highlights the mismatch in the referral and counter-referral system. The difficulty in getting these people into Primary Health Care may be due to the invisibility that the disease has had over the years in the health system[13]. It is important to note that this fact is not exclusive to Brazil14. It can also be seen that official bodies themselves, unintentionally, can contribute to maintaining the situation. One example is the document published in 2018 by the Brazilian Ministry of Health, entitled Clinical Protocol and Therapeutic Guidelines for Sickle Cell Disease[15], which does not include the treatment of ulcers in its content.

In 2006, in order to promote the inclusion of these individuals in Primary Health Care, the Ministry of Health launched the sickle cell disease manual for Community Health Worker[16]. With regard to ulcers, the document recommends routine dressing and rest in the acute phase with elevated limbs. Therefore, it does not highlight the importance of the person being monitored by a nurse from the Primary Care Center or specialized service with knowledge and skills in ulcer treatment.

The Community Health Worker t is responsible for making at least one monthly visit to each family[15]. However, the results of the study revealed that this practice is not effectively applied, since only half of the people with ulcers reported having received at least one monthly visit from this professional. This discrepancy is relevant data that instigates discussion about the effectiveness of the network and the performance of the different actors within it. In addition, previous studies carried out in regions in the interior of Minas Gerais corroborate this need. This study indicated that home visits do not follow a systematized approach, and professionals are often unaware of the specifics of monitoring people with sickle cell disease[17].

In the United States, care is usually provided by a hematologist in collaboration with the primary care physician. In some cases, a single setting is used, such as a comprehensive sickle cell clinic. Often, a primary care doctor with expertise in treating sickle cell disease acts as the sole provider. Referral to other specialists is made according to the need to control the complications of the disease[18].

The Brazilian Ministry of Health published a manual[19] in 2012 aimed at guiding professionals in the Unified Health System (*Sistema Único de Saúde -SUS*) at all levels of care. Later, in 2015, another document was released[13], which defined the Family Health Team as responsible for promoting self-care in the prevention of leg ulcers, as well as following the guidelines for treatment. However, despite the current guidelines, the study found that 66.7% of people with ulcers in Minas Gerais were treated exclusively at home, and only 5.6% received ongoing care in Primary Health Care. This data shows the limited accessibility of this population to this level of care. This can be partly explained by the perceptions of community workers. They believe that primary health care professionals are not adequately prepared in terms of knowledge, skills and attitudes to care for people with sickle cell disease[17].

A study carried out in the United States analyzed data from the OneFlorida Data Trust health system. This is a centralized repository of electronic medical record data from eight different health systems in Florida. The number of adults included with sickle cell disease was 1,147. The majority of patients were seen only by a primary care provider (30.4%), followed by primary care providers and a hematologist (27.5%), neither primary care providers nor a hematologist (23.3%) and only a hematologist (18.7%). The authors concluded that patients with sickle cell disease who have both a primary care provider and a hematologist involved in their care benefit from fewer hospitalizations[20].

Brazilian nurses working in Primary Health Care have misconceptions about sickle cell disease. In addition, there is a lack of effective follow-up of patients within the area covered by these professionals, resulting in a failure to establish a bond between the patient and the Primary Care team. This gap reflects the discrepancy between the recommendations for care and the practice observed in Primary Health Care[21]. In order to face these challenges, it is crucial to implement strategies that promote the updating of nursing professionals' knowledge and the improvement of care processes, with the aim of guaranteeing effective care centered on patients' needs.

International literature confirms the fragmented and inadequate management of people with sickle cell disease and leg ulcers. Often, this responsibility falls to primary care professionals, who sometimes lack the necessary knowledge to meet the care demands required[22]. India recommends that the primary health care system be strengthened to screen and manage people with sickle cell disease with appropriate community mobilization activities. The program should establish partnerships with traditional healers and community

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leaders, taking into account the cultural specificities of the country. People should be encouraged to seek treatment[23].

Primary health care teams are instructed to refer individuals to specialized care if the wounds do not regress after 2 months of treatment[19]. However, this guidance often results in discontinuity of care, due to the lack of specialized services and qualified professionals to treat this condition in Minas Gerais.

In many services, Primary Health Care does not have an established protocol for the care of patients with sickle cell ulcers, and lacks a systematic approach. Care is conducted in a reactive manner, with the patient seeking assistance when necessary. Dressings are changed at home, with periodic assessments taking place in primary care. This leads patients to visit the health center only occasionally. In addition, the absence of nurses in this process is notable. This often leads to the transfer of care to nursing technicians, without the supervision of the professional nurse[24].

Another challenge in the treatment of wounds in Primary Health Care is the lack of physical and material resources, which are often technologically outdated, as well as the shortage of qualified human resources[25].

In the study carried out, the most common topical treatments were ointments with collagenase or antibiotics and interactive coverings. It's important to note that in some of the patients, the treatment was chosen by the patient themselves, family members or friends. In almost half of the cases, doctors were the prescribers of the topical treatment, most of them indicating the ointment. When nurses were responsible for the prescription, the treatment indicated fell on interactive coverings. This highlights the responsibility of nurses to assess, prescribe and perform dressings for different types of wounds, as well as to manage edema in the lower limbs by applying inelastic and elastic compression therapy when the ulcer is diagnosed[26].

The findings on topical treatment are divergent when compared with data from a specialized service staffed by stomatherapist nurses[26]. The treatment of sickle cell disease ulcers remains a challenge. New studies are underway to evaluate the efficacy of topical treatments and describe the ulcer microbiome[27]. However, it is well known that occlusive dressings are essential for wound healing to ensure a favorable microenvironment for healing[26].

Adequate local care for the ulcer includes debridement, control of the bacterial load or infection and maintenance of the temperature and moist environment in the wound bed, which are achieved by means of occlusive dressings[22]. However, the current study showed that most participants use topical treatments that don't promote occlusion and require daily changes. Furthermore, the Ministry of Health's manual[19] recommends adjuvants (hydrogels), essential fatty acids, ointments that promote enzymatic debridement and antibiotic ointments. In addition, it proposes hyperbaric oxygen therapy and vacuum therapy for ulcers resulting from sickle cell disease[19], although the benefits have yet to be proven[27]. The inconsistency of current recommendations in Ministry of Health documents confirms the need to update and periodically review institutional protocols, in particular updating the Sickle Cell Disease/Ulcers: Prevention and Treatment Manual[19] with evidence-based recommendations to guide the practice of nurses who care for people with ulcers. In addition, it is imperative to update the Clinical Protocol and Therapeutic Guidelines for Sickle Cell Disease[15] to include specific inputs for the treatment of people with leg ulcers.

Venous insufficiency is frequently observed in people with sickle cell disease, highlighting the importance of not only treating the ulcer topically, but also managing leg edema through containment or compression therapies - the latter being the preferred approach[22]. In this study, around 76.4% of the participants had edema, while only 11% used compression therapy, and of these, 7.3% used the Unna boot.

An average cost of R$28.33 was calculated for each dressing change in the Stomatherapy Service of a public health institution, including the nursing consultation, ulcer cleaning materials, interactive dressings and the Unna boot to treat edema[26]. Notably, treatment with interactive dressings, which require fewer weekly changes, has a lower cost compared to traditional dressings[28]. Furthermore, it is essential to consider the importance of addressing the underlying pathophysiology of the condition[4].

Regarding to the supply of the materials needed to treat ulcers, it was found that almost half of the participants bought all the materials themselves. This has an impact on the financial budget and can have repercussions on patients' lives. The federal or state level should provide specific financial resources to pay for the treatment of patients with ulcers.

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It is important to establish a network of care for sickle cell disease patients with leg ulcers, as this has not been identified in Minas Gerais. In addition, the professionals belonging to this network need to have knowledge of the etiology of the ulcer, its chronicity, the healing mechanisms and the factors that affect it, as well as knowledge of the products used in wound treatment. These requirements are fundamental to inform clinical decision-making[29].

The study's findings highlight the lack of specialized units for monitoring and treating patients with sickle cell disease and ulcers. This reveals the invisibility of this group in the formulation of public policies aimed at assisting people with leg ulcers. Thus, considering the particularities and complexity of wound care[29], it is crucial to reorganize health services, including the implementation of specialized wound care units. This is essential to meet the specific needs of this population and ensure continuity of care.

Limitations of the study

Some limitations should be considered when interpreting the results of this study. Firstly, the Hemominas Foundation's electronic medical records system did not allow for the identification of patients with leg ulcers. Searching for other solutions for identification could introduce a selection bias and possibly result in not identifying all the people with active ulcers registered with the foundation. It is important to note that interviews were used to collect the data, which could lead to memory bias on the part of the participants.

CONCLUSION

Most of the participants were not included in the Health Care Network for ulcer treatment and did not receive systematized care or appropriate supplies. Most of the individuals received care at home, in Secondary Care, and had periodic follow-up with hematologists at blood centers.

The findings identify the need for specific public policies for people with sickle cell disease and leg ulcers, with the allocation of financial resources. It is crucial to review the organization of the Health Care Network so that it is able to accommodate these people. The study highlights the need to review current documents published by the Ministry of Health for the more comprehensive and integrated management of leg ulcers in patients with sickle cell disease, stimulating future research and interventions that address the gaps identified.

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Authors' contributions

Conceptualization, E.L.B. and J.A.O.S.; Methodology, E.L.B. and J.A.O.S.; Validation, E.L.B. and J.A.O.S.; Formal Analysis, E.L.B., J.A.O.S., P.G.R.A., C.R.L. and M.L.M.S.; Investigation, E.L.B. and J.A.O.S; Data Curation, E.L.B., J.A.O.S. and P.G.R.A.; Manuscript Writing, E.L.B., J.A.O.S., P.G.R.A., C.R.L. and M.L.M.S.; Writing – Review and Editing, E.L.B., J.A.O.S., P.G.R.A., C.R.L. and M.L.M.S.; Visualization, E.L.B., J.A.O.S., P.G.R.A., C.R.L. and M.L.M.S.; Supervision,E.L.B.; Project Administration, E.L.B. and J.A.O.S. All authors read and agreed with the published version of the manuscript.