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Review Article

ASPECTOS SOCIAIS DOS PACIENTES COM ÚLCERA DE PERNA NA DOENÇA FALCIFORME: REVISÃO INTEGRATIVA

SOCIAL ASPECTS OF PATIENTS WITH LEG ULCER IN SICKLE CELL DISEASE: INTEGRATIVE REVIEW

ASPECTOS SOCIALES DE LOS PACIENTES CON ÚLCERA DE PIERNA EN LA ENFERMEDAD FALCIFORME: REVISIÓN INTEGRADORA

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RESUMO

Objetivo: identificar à luz da literatura como o aspecto social interfere na vida do paciente com úlcera de perna proveniente da Doença Falciforme (DF). **Método:** trata-se de uma revisão integrativa, baseada nas fontes de dados U.S. National Library of Medicine - NCBI (PUBMED), Biblioteca Virtual em Saúde (BVS), Literatura Latino-americana em Ciências da Saúde (LILACS), Scientific Electronic Library Online (SCIELO), Medical Literature Analysis and Retrieval System Online (MEDLINE) e Base de Dados em Enfermagem (BDENF), no período de agosto de 2015 a junho de 2016. A partir dos critérios de inclusão e exclusão, foram selecionados 11 artigos. **Resultados:** o contexto social e racial em que os indivíduos com DF estão inseridos propicia condições socioeconômicas e educacionais desfavoráveis; aliados a esses problemas surgem as complicações da doença como a úlcera de perna que influencia negativamente na vida desses pacientes e geram limitações físicas e laborais. **Conclusão:** esta revisão forneceu um panorama de como o contexto social impacta na vida desses pacientes, assim, julga-se necessário um delineamento de ações e iniciativas de políticas públicas de saúde com recorte racial no intuito de promover saúde e assegurar a equidade, um dos princípios do Sistema Único de Saúde.
Descritores: Anemia falciforme; Úlcera da perna; Fatores socioeconômicos; Estigma social.

ABSTRACT

Objective: to identify the light of literature as the social aspect interferes in the patients life with leg ulcer from the Sickle Cell Disease (SCD). **Method:** this is an integrative review, based on data sources US National Library of Medicine - NCBI (PUBMED), Virtual Health Library (VHL), Health Sciences Latin American Literature (LILACS), Scientific Electronic Library Online (SCIELO), Medical Literature Analysis and Retrieval System Online (MEDLINE) and Nursing Database (BDENF), from August 2015 to June 2016. Starting from the inclusion and exclusion criteria, 11 articles were selected. **Results:** the racial and social context in which individuals with SCD are inserted, provides socioeconomic and educational conditions unfavorable; Allied to these problems arise complications of the disease such as leg ulcers which negatively influences in the patient's lives and generate physical and labor constraints. **Conclusion:** this review provided an overview of how the social context impacts on the lives of these patients, so, it is deemed necessary an outline of actions and public health policy initiatives with cutting racial in order to promote health and ensuring equity, one of the Unified Health System principles.
Descriptors: Anemia, sickle cell; Leg ulcer; Socioeconomic factors; Social stigma.

RESUMEN

Objetivo: identificar a la luz de la literatura como el aspecto social interfiere en la vida del paciente con úlcera de pierna proveniente de la enfermedad Falciforme. **Método:** se trata de una revisión integradora, basada en las fuentes de datos U.S. National Library of Medicine - NCBI (PUBMED), Biblioteca Virtual en salud (BVS), literatura Latinoamericana en Ciencias de la Salud (LILACS) Scientific Electronic Library Online (SCIELO), Medical Literature Analysis and Retrieval System Online (MEDLINE) y base de datos en Enfermería (BDENF), en el periodo de agosto de 2015 a junio de 2016. A partir de los criterios de inclusión y exclusión, fueron seleccionados 11 artículos. **Resultados:** el contexto social y racial en el que los individuos con enfermedad Falciforme están insertados, propicia condiciones socioeconómicas y educacionales desfavorables; aliados a esos problemas surgen las complicaciones de la enfermedad como la úlcera de pierna que influencia negativamente en la vida de esos pacientes y generan limitaciones físicas y laborales. **Conclusión:** esta revisión proveyó un panorama de cómo el contexto social impacta en la vida de esos pacientes, así, se juzga necesario un delineamiento de acciones e iniciativas de políticas públicas de salud com recorte racial con la finalidad de promover salud y asegurar la equidad, uno de los principios del Sistema Único de Salud.
Descriptores: Anemia de células falciforme; Úlcera de la pierna; Factores socioeconómicos; Estigma social.

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INTRODUCTION

Sickle Cell Disease (SCD) is considered to be a serious global public health problem, predominantly in the black population, because it has an impact on the morbidity and mortality of this population[1-2]. In Brazil, it is considered as the most commonly found hematological genetic disorder. It results from a change in hemoglobin (Hb) due to the substitution of glutamic acid for valine at the 6-position of the hemoglobin β chain, which results in a production of mutant hemoglobin (HbS). Thus, the red cell is no longer rounded and acquires the shape of sickle, hence the sickle's name[3-4].

According to the World Health Organization (WHO), approximately 300,000 children are born with SCD in the world each year, with high incidence in Africa, Saudi Arabia and India. In Brazil, the highest incidences are found in Bahia (1:650), Rio de Janeiro (1:1300) and in the States of Pernambuco, Maranhão and Minas Gerais (1:1400), due to the large Afrodescendant population[5-7]. In relation to the Brazilian prevalence, the North and Northeast regions present the highest rates (6 to 10%)[8]. Statistical data presented by the National Neonatal Screening Program (PNTN) of the Ministry of Health indicate that 3,500 children / year with SCD and 200,000 with sickle cell trait are born in Brazil, and it is estimated that 7,200,000 people carry the sickle cell trait (HbAS) and between 25,000 and 30,000 with SCD[1]. In this perspective, it is believed that more than 80% of these births occur in low or middle-income countries[9].

The SCD lethality is 80% in children under 5 years of age who do not receive the necessary health care. Meanwhile, the average life of people with SCD reaches the age of 48 years[5]. Therefore, because it represents a high degree of suffering for the patient, the early diagnosis, treatment and care plan contributes to the patient enjoying a longer and better quality of life[10].

In recognition of the problem, in 2001, the Ministry of Health, through Ordinance No. 822/01, included the examination that detects SCD and other hemoglobinopathies in the PNTN[1]. Subsequently, in order to modify the natural history of SCD in Brazil, promote longevity with quality of life and guide people with sickness or sickle cell trait, the Ministry of Health created on August 16, 2005 the National Policy of Integral Attention nº 1391, with the purpose of promoting health, preventing complications, performing an early diagnosis, treating, rehabilitating health problems and articulating the technical areas whose actions are interfaced with haematological and hematotherapeutic care[11].

The main mechanisms of the pathophysiology of SCD are related to genetic factors and to vessel obstructions, especially in the microvasculature, since the sick red blood cells cause vaso-occlusion, alter the concentration of fetal hemoglobin and inhibit the nitric oxide synthase enzyme (NOS). Thus, one of the main clinical manifestations of the disease is the leg ulcer and the factors influenced by this problem, such as psychosocial, emotional and economic changes, due to the chronicity of the lesions, the high rates of recurrence accompanied by intense pain and sometimes secretion and sharp odor, as well as the difficulty of healing that implies in compromise of the quality of life of patients with SCD[2,12].

Therefore, this study is based on the assumption that sickle cell patients with leg ulcers are predisposed to confront the disease with difficulty, given the great social impact related to this problem. Thus, the objective is to identify, in the light of the literature, how the social aspect interferes in the life of the patient with leg ulcer coming from the SCD.

METHODS

This is an integrative bibliographical review, which consists of a broad methodological approach regarding revisions, with the purpose of analyzing all literature on a delimited topic, for being able to synthesize and comprehend in a more complete way the analyzed phenomenon [13-14].

Integrative review differs from other review methods because it allows greater reliability by describing previous studies to support Evidence-Based Practice (EBP) and provide an arsenal of knowledge that will serve as a foundation for improvements in patient care. The use of this methodology provides a comprehensible overview of complex concepts, theories and problems relevant to health professionals, not only for the development of protocols, procedures and public policies, but also for stimulating critical thinking necessary for daily

professional practices in order to improve the quality of patient care[13,15].

For the elaboration of the study, six stages were covered: identification of the problem and definition of the guiding question; search in the literature and construction of inclusion and exclusion criteria; data collection and categorization of studies; critical analysis of included studies; discussion of results; and presentation of the integrative review[14]. For this, it was necessary that the guiding question and the objective were well elaborated for the construction of the research, as well as the results of the studies were synthesized and critically analyzed[13]. Thus, the first part of the research was the elaboration of the guiding question that consisted in: "How does the patient with leg ulcer, triggered by Sickle Cell Disease, experience the problems related to the social context?". The search and sampling phase in the literature, carried out from August 2015 to June 2016, was carried out through the online access of the U.S. National Library of Medicine - NCBI (PUBMED) and the Virtual Health Library (BVS), specifically in the following electronic databases: Latin American Literature in Health Sciences (LILACS), Scientific Electronic Library Online (SCIELO), Medical Literature Analysis and Retrieval System Online (MEDLINE) and Database of Nursing (BDENF). The descriptors were selected from the terminology in health consulted in the Descriptors in Health Sciences (DeCS), in the Portuguese, English and Spanish languages, being the following: Anemia Falciforme/ *Anemia, Sickle Cell/ Anemia de Células Falciformes;* Úlcera da Perna/ *Leg Ulcer/ Úlcera de la Pierna;* Fatores Socioeconômicos/ *Socioeconomic Factors/ Factores Socioeconómicos;* Estigma Social/ *Social Stigma/ Estigma Social.* Considering the difference between the terms Sickle Cell Anemia (SCA) and Sickle Cell Disease, which encompasses a group of hematological disorders of genetic origin, the descriptor was chosen due to methodological rigor in the research area.

Descriptors were cross-referenced in all databases using the Boolean operator *AND.* Thus, the associations with the descriptors were association 01: "Anemia Falciforme" *and "*Úlcera da Perna"; *"Anemia, Sickle Cell" and "Leg Ulcer"; "Anemia de Células Falciformes" and "Úlcera de la Pierna"*; association 02: "Anemia Falciforme" *and "*Fatores Socioeconômicos"; *"Anemia, Sickle Cell" and "Socioeconomic Factors"; "Anemia de Células Falciformes" and "Factores Socioeconómicos"*; association 03: "Anemia Falciforme" *and "*Estigma Social"; *"Anemia, Sickle Cell" and "Social Stigma"*; *"Anemia de Células Falciformes" and "Estigma Social".*

The following articles were included in relation to the associations: scientific articles that addressed the guiding question, published in full in online format, in national, international and free journals available in Portuguese, English, and Spanish and published in the last ten years (2005 to 2015). Publications that were not made available online were excluded, as well as articles not indexed in the *Qualis* platform and repeated articles in different databases.

According to research on the VHL platform, association 01 resulted in 222 articles, association 02: 165 articles and the association 03: 24 articles. In the *PubMed* database, association 01 obtained a total of 225 articles, association 02: 330 articles and association 03 a total of 15 articles. After applying the inclusion and exclusion criteria from the reading of the titles and abstracts, the sample was of 19 articles in the BVS and 2 articles in the *PubMed* (Table 01).

Table 1 – Summary table of the selected samples in the BVS and *PubMed* electronic databases.

ASSOCIATIONS	**ASSOCIATION 01**		**ASSOCIATION 02**		**ASSOCIATION 03**	
	BVS	*PubMed*	BVS	*PubMed*	BVS	*PubMed*
Studies Found	222	225	165	330	24	15
Available	59	50	80	87	9	5
Year of Publication (2005 - 2015)	46	28	63	55	8	4
Language (English)	39	28	59	55	6	3

Language (Portuguese)	8	0	3	0	1	0
Language (Spanish)	0	0	0	0	0	0
Exclusions	2	21	2	27	0	2
Sample	45	7	60	28	7	1
Total of Samples	8	0	9	2	2	0

Source: The authors

In view of the above, a full reading of the selected articles was carried out. Data were extracted by means of a data collection instrument adapted from Ursi[14] composed of the following items: year, author(s), periodical and title, as well as a spreadsheet developed by the authors in the *Excel* as a way to organize the information and make it easy to access and manage.

The critical analysis of the studies occurred from a detailed and interpretative evaluation in order to look for different and conflicting results in the selected articles, in order to include the most important ideas for the development of a consistent thought within the proposed theme.

For the classification of evidence of the studies, the proposal of the *Agency for Health care Research and Quality* (AHRQ) was used, considering seven levels of evidence: 1. Meta-analysis of multiple controlled studies; 2. Well-delineated randomized controlled clinical trial; 3. Well-delineated clinical trials without randomization; 4. Well-designed cohort and case-control studies; 5. Systematic review of descriptive and qualitative studies; 6. Descriptive or qualitative study; 7. Opinion of authorities and/or expert committees report[16].

RESULTADOS E DISCUSSÃO

A interpretação dos resultados foi baseada nos dados literários com a finalidade de comparar os dados contidos a partir das análises das publicações. Dos 21 artigos selecionados nas bases de dados consultadas foram excluídos 10 por não responderem à pergunta norteadora, sendo assim a amostra final foi de 11 artigos. Com o desígnio de uma maior compreensão da busca bibliográfica realizada e categorização dos estudos, foi elaborado um quadro sinóptico (Figura 01).

Figure 1 – Selected articles for synthesis of the integrative review, according to year, author(s), periodical and title.

Year	Author(s)	Journal	Title
2013	Martins A, Moreira DG, Nascimento EM, Soares E [17]	Esc. Anna Nery	Self-care for the treatment of sickle leg ulcer: nursing guidelines
2015	Alencar SS, Junior CJC,Guimarães BF, Cunha DP, Rocha LV, Teixeira FEN, Oliveira CDL [18]	Rev. med. Minas Gerais	Most prevalent clinical complications in patients with Sickle Cell Disease in a medium-sized city in Minas Gerais, Brazil
2005	McClish DK, Penberthy LT, Bovbjerg VE, Roberts JD, Aisiku IP, Levenson JL, Roseff SD, Smith WR [19]	Health and Quality of Life Outcomes	Health related quality of life in sickle cell patients: the PiSCES project
2013	Santos JP, Gomes NM [20]	Rev. bras. hematol. hemoter.	Sociodemographic aspects and quality of life of patients with sickle cell anemia
2013	Pereira SAS, Brener S, Cardoso CS, Proietti ABFC [21]	Rev. bras. hematol. hemoter.	Sickle Cell Disease: quality of life in patients with hemoglobin SS and SC disorders
2010	Roberti MRF, Nunes CS, Moreira SO, Tavares RS, Filho HMB, Silva AG, Maia CHG, Lima FL, Teixeira DF Reciputti BP, Filho CRS, Filho JS, Santos DB, Lemos IP [22]	Rev. bras. hematol. hemoter.	Evaluation of the quality of life of patients with Sickle Cell Disease in a General Hospital of Goiás, Brazil
2015	Fernandes TA, Medeiros TM, Alves JJ, Bezerra CM, Fernandes JV, Serafim ES, Fernandes MZ, Sonati MF [23]	Rev. bras. hematol. hemoter.	Socioeconomic and demographic characteristics of sickle cell disease patients from a low-income region of northeastern Brazil

2008	Pereira SAS, Cardoso CS, Brener S, Proietti ABFC [24]	Rev. bras. hematol. hemoter.	Sickle cell disease and quality of life: a study of patients' subjective perception of Hemominas Foundation, Minas Gerais, Brazil
2014	Haywood CJ, Lanzkron S, Bediako S, Strouse JJ, Haythornthwaite J, Carroll CP, Diener-West M, Onojobi G, Beach MC [25]	J Gen Intern Med	Perceived discrimination, patient trust, and adherence to medical recommendations among persons with sickle cell disease
2010	Felix AA, Souza HM, Ribeiro SB [26]	Rev. bras. hematol. hemoter.	Epidemiological and social aspects of Sickle Cell Disease
2013	Silva HD, Paixão GPN, Silva CS, Bittencourt IS, Evangelista TJ, Silva RS [27]	rev. cuid.	Sickle Cell Anemia and its psychosocial aspects: the gaze of the patient and the family caregiver

Source: The authors.

Of the selected articles, five (45.5%) had level of evidence as 5, four (36.6%) with level of evidence as 4, one (8.9%) with level of evidence as 2 and one (8.9%), with level of evidence as 1. Most of the articles have level of evidence as 4 and 5, as it results from well-delineated cohort, case-control, and systematic descriptive and qualitative review due to the type of research line practiced in this study, according to the AHRQ[16].

The studies were published in separate journals of medical and public health, whose *Qualis* ranged from B1 to B5 (Figure 02).

Figure 2 – Classification of articles according to level of evidence, *Qualis* periodicals, lead author and origin.

Association	Title	Level of Evidence	Qualis Journals	Main author	Origin
Association 01	Self-care for the treatment of sickle leg ulcer: nursing guidelines	3	B1	Nurse	Brazil
	Most prevalent clinical complications in patients with Sickle Cell Disease in a medium-sized city in Minas Gerais, Brazil	5	B5	Medical Academic	Brazil
Association 02	Health related quality of life in sickle cell patients: the PiSCES project	4	B1	Medical	United States
	Sociodemographic aspects and quality of life of patients with sickle cell anemia	4	B4	Physiotherapist	Brazil
	Sickle Cell Disease: quality of life in patients with hemoglobin SS and SC disorders	2	B4	Pedagogue	Brazil
	Evaluation of the quality of life of patients with Sickle Cell Disease in a General Hospital of Goiás, Brazil	4	B4	Medical	Brazil
	Socioeconomic and demographic characteristics of sickle cell disease patients from a low-income region of northeastern Brazil	5	B4	Pharmacist	Brazil
	Sickle cell disease and quality of life: a study of patients' subjective perception of Hemominas Foundation, Minas Gerais, Brazil	5	B4	Pedagogue	Brazil
	Perceived discrimination, patient trust, and adherence to medical recommendations among persons with sickle cell disease	4	B1	Medical	United States
	Epidemiological and social aspects of Sickle Cell Disease	5	B4	Nurse	Brazil

Association 03	Sickle Cell Anemia and its psychosocial aspects: the gaze of the patient and the family caregiver	5	B5	Nurse	Brazil

Fonte: Os autores.

The SCA is a chronic disease of high influence on self-esteem, in the formation of personality and in the construction and maintenance of family and social relationships. These characteristics encourage a feeling of punishment and cause the patient to experience feelings of sadness, contempt, anguish, impotence and uselessness. These thoughts and judgments provide a self-exclusion, both for diagnosis and for the ethnic origin of the disease, followed by prejudice and racial discrimination, still so crystallized in our society, and by the negative impact on the patients' lives, sickness and death[27].

When analyzing the racial issue with greater amplitude, studies indicate that the individuals with SCD are predominantly of the black race and have low schooling. In this context, the Institute of Applied Economic Research (IPEA)[28] states that gender and race inequalities are contained in Brazilian society and limit access, progression and opportunities for the black population[18,22 -24].

Discussions about the labor market deserve to be highlighted as being a crucial factor for the construction of personal, social and, above all, economic identity, in order to guarantee survival. Individuals with SCD, of economically active age, have basic schooling and, consequently, low remuneration of up to two minimum wages. The complications arising from the disease result in lower professional competitiveness and lower purchasing power in order to influence negatively on these people's lives[18,20-21,23-24,26].

As for school, it is possible that in some moments the patient needs to be absent due to episodes of pain and the need for constant outpatient follow-up and consultations and examinations. This absence of schooling can jeopardize the educational development and justify the socioeconomic problems of SCD patients[19]. However, a study that evaluates the quality of life of sickle cell patients has resulted in the fact that even patients with low educational level and being victims of prejudice due to SCD, the majority of the subjects evaluated in a positive way both their quality of life and their health[22].

In addition to socioeconomic problems, individuals with SCA experience various complications of the disease from physiopathological mechanisms, such as leg ulcers, which appear more frequently in patients living in tropical areas (50% higher than in other regions), male (2M:1F) and aged between 10 and 50 years. They may be single or multiple lesions that are generally shallow, with raised borders, presence of exudate and necrotic material at the base of the lesion, and hyperpigmentation in peri-lesion region with loss of hair follicles.

They occur frequently in the lower limbs, in the areas with lower subcutaneous tissue and presence of thin skin, such as the internal and external malleolar region, the anterior tibial area, the Achilles tendon area and, to a lesser extent, the dorsum of the foot. The etiology can be traumatic due to insect bites or spontaneous; they cause painful moments and long periods of treatment due to the high rates of recurrence (25% to 97%) and slow healing, in addition to burdening health services[4,17-18].

When considering that patients with SCA compared to the general population are ten times more likely to develop leg ulcers, it is essential to maintain general measures and treatment sites, such as self-care, trauma prevention and dressing. It should be noted that leg ulcers strongly interfere with patients' lives and determine their lifestyle, since the limitations are associated with coexistence with severe pain, loss of autonomy to perform daily activities and decrease of mobility, which conditions a physical disability[4,17-18].

It is relevant to point out the existence of a parallel between discrimination and non-adherence to medical recommendations in people diagnosed with SCA. A study shows that adequate communication between the target audience and health professionals can facilitate adherence to treatment; on the other hand, the lack of a link and a welcoming dialogue promotes insecurity in patients and leads to abandonment of treatment and failure to carry out self-care[25].

FINAL CONSIDERATIONS

Because it is a chronic disease, of a hereditary nature and irreversible, the patient with SCD encounters difficulties mainly when exposed to clinical manifestations, such as leg ulcers, which causes a change in their daily life due to the limitation and the difficulty to face the disease.

This leads us to observe that individuals with SCD are doubly affected, both by the stigma of the disease and by the social context in which they are inserted. Thus, it is considered necessary to outline actions and initiatives of public health policies with racial cut-off in order to promote health and ensure equity, one of the principles of the Unified Health System.

When considering the obstacles of a social and / or economic nature faced by patients with SCD and the magnitude of the negative impact of this issue, the health professional needs to sensitize and understand the perspective of these subjects to be able to develop educational actions directed to the individual and collective needs of this population. In addition, it is essential to include strategies that offer self-knowledge, self-control and active participation in treatment, making them autonomous protagonists of their health, capable of transforming the natural history of the disease into a process of longevity.

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