ISSN: 1676-4285

OBJN Online Brazilian Journal of Nursing
ENGLISH
Federal Fluminense University
AURORA DE AFONSO COSTA NURSING SCHOOL
uff
Review Articles

Families of children with sickle cell disease: an integrative review

Elaine Cristina Rodrigues Gesteira[1], Regina Szylit Bousso[2], Maira Deguer Misko[3], Carolliny Rossi de Faria Ichikawa[2], Patrícia Peres de Oliveira[1]

1 Federal University of São João del-Rei
2 São Paulo University
3 Federal University of São Carlos

ABSTRACT

Aim: To identify scientific evidence about families of children with sickle cell disease. **Method:** An integrative review of the search used the following descriptors: anemia sickle cell; hemoglobin SC disease; hemoglobin sickle; family; family relations; and, child. The search was carried out for literature in Portuguese, English, and Spanish. The search used databases such as MEDLINE, LILACS, CINAHL, and PubMed, and looked for articles published between January 2005 and January 2015. **Results:** There were 16 articles selected that originated the following thematic categories: *the impact of sickle cell disease on the family*, *psychosocial effects on family dynamics*, *child's quality of life*, and *family and social support/support networks*. **Discussion:** The studies report that the family remains the main provider of care for children with sickle cell disease, and that families faces challenges in achieving comprehensive care and fighting for their children's quality of life. **Conclusion:** This evidence will provide support for multi-professional teams in the construction of continuous care for the families of children who are sickle cell patients.

Descriptors: Anemia, Sickle Cell; Hemoglobin, Sickle; Family; Pediatric Nursing.

Gesteira ECR, Bousso RS, Misko MD, Ichikawa CRF, Oliveira PP. Families of children with sickle cell disease: an integrative review. Online braz j nurs [internet] 2016 Jun [cited year month day]; 15 (2):276-290. Available from: http://www.objnursing.uff.br/index.php/nursing/article/view/5289

INTRODUCTION

Sickle cell disease (SCD) belongs to a group of the most common genetic disorders in Brazil, caused by modified hemoglobin (HbS). Generally, parents are asymptomatic of a single affected gene (heterozygous) and produce HbA and HbS, communicating it to the child who receives the modified gene in a double dose (HbSS) featuring sickle cell anemia, one of the most severe forms of hemoglobinopathies.

The presence of S variant hemoglobin in red blood cells impairs normal functioning, causing a decrease in the survival of erythrocytes (hemolysis) and, consequently, symptoms related to vaso-occlusive phenomena[2].

The genesis of the disease occurred in Africa and spread to the Americas through slavery. Thus, there has been an increase in Brazil's Afro-descendant population. SCD constitutes a public health problem because of its prevalence and clinical importance[3-4].

Clinical manifestations of SCD start around the sixth month of a child's life and include painful crises, jaundice, chronic anemia, priapism, increased risk for cerebrovascular accidents (CVA), gallstones, and recurrent infections, especially of the respiratory tract[5].

The presence of physical symptoms mainly relates to painful reactions, which interfere with the daily life of children with SCD. This pain also extends to the child's family because, often, the family has not been prepared and they are not ready to face the illness and bear the anguish of their relative. In the case of diagnosis of SCD, the effect is even more devastating[6-7] due to the need for pain management and the complexity of the treatment, the disease's emotional and psychosocial impact, and the fact that the family is, at present, the primary source of support for affected individuals. Thus, when any loved one presents risks through any imbalance, the family needs to adapt to the new situation[7-8].

In this sense, the child's family takes on the continuum of care, which is mainly related to the worsening of symptoms and the monitoring of painful crises[9].

Currently, few studies explore families' experiences of SCD childcare. Most studies examine the biological, pharmacological, and genetic aspects of the disease.

Thus, this study is justified by the need to investigate scientific evidence about the subject, gathering results that promote nursing interventions for families of children with SCD.

In doing so, it is believed that health professionals are given the opportunity to reflect on the issue, thus sensitizing them to the planning and implementation of effective actions that can ensure a better quality of life for children with SCD and their families.

Therefore, the aim of this study is to identify scientific evidence about families of children with SCD.

METHOD

This is a study that uses the integrative review, a comprehensive method that simultaneously includes experimental and non-experimental research to achieve a more complete understanding in terms of a phenomenon of interest by presenting the current state of science and its applicability to nursing practice[10].

For the preparation of this integrative review, the following steps were taken: the identification of the problem (clear definition of the purpose of the review), a literature

search (delimiting keywords, databases, and the establishment of inclusion and exclusion criteria for selecting articles), and assessment and data analysis[10].

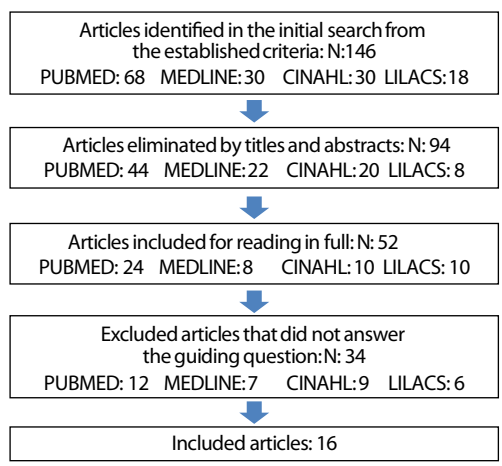
To guide the integrative review, the following question was formulated: what is the scientific evidence in relation to families with children suffering from SCD?

By using the electronic databases MEDLINE (International Literature on Health Sciences), LILACS (Latin American and Caribbean Health Sciences), CINAHL (Cumulative Index to Nursing and Allied Health Literature), and PUBMED (National Library of Medicine and National Institutes of Health), the authors independently conducted the survey. The aim was to ensure the legitimacy of the study; the online search of scientific articles only looked for those published in the last 10 years, that is, from January 2005 to January 2015, due to the consolidation of the *Programa Nacional de Triagem Neonatal* (PNTN) (National Neonatal Screening Program) for hemoglobinopathies, which began in 2005 since Ordinance GM/MS 822 of 06.06.2001. It took four years to consolidate PNTN throughout Brazil. It is also worth mentioning that in other countries, such as Britain and the U.S., neonatal screening for SCD was mandatory from 2006, the same year that the World Health Organization (WHO) recognized SCD as a public health problem in Africa[3].

Inclusion criteria were: articles published in Portuguese, English, or Spanish, with abstracts available in the databases selected in the period 2005-2015; published articles whose methodology includes family participants or parents, caregivers, and children with SCD; and articles that examine aspects of family life regarding SCD and care for those affected. The search excluded articles that did not address the research question, as well as those whose summary was not in the databases, and those that were not available in full text format or written in a language other than those defined in the study. The search also excluded abstracts and annals of congresses, reviews, editorials, opinions, previous notes, reports, articles of narrative review, and critical review.

To perform the search, combinations were used between keywords: descriptors in DeCS (health sciences descriptors) and MeSH (medical subject headings "anemia sickle cell"), [MeSH terms] OR "hemoglobin SC disease" [DeCS terms] OR "hemoglobin, sickle" [DeCS terms] AND "family" [MeSH terms] OR "family" [DeCS terms] OR "family relations" [MeSH terms] OR "family relations" [DeCS terms] AND "child" [MeSH terms] OR "child" [DeCS terms]. In CINAHL databases and portals, PUBMED descriptors in English were used; for accessing the MEDLINE database, descriptors in Portuguese, Spanish, and English were used; in the LILACS database, the descriptors used were in Portuguese and Spanish. From the combination of these descriptors, 146 references were found, as shown in **Figure 1**.

Figure 1 - Flowchart for the selection of articles. Divinópolis, 2015.



Gesteira ECR, Bousso RS, Misko MD, Ichikawa CRF, Oliveira PP. Families of children with sickle cell disease: an integrative review. Online braz j nurs [internet] 2016 Jun [cited year month day]; 15 (2):276-290. Available from: http://www.objnursing.uff.br/index.php/nursing/article/view/5289

Afterwards, an analytic reading of these studies (16 articles) was performed. The articles were found and numbered in order of location, and the data were organized by the meaning of the information to be extracted from elected posts. The core of the integrative review is the categorization of studies. In this regard, the procedure is the presentation of results and the descriptive discussion of information through the construction of a summary table containing: a study code according to the classification of the authors; goals; journal; year of publication; study characteristics; and another synoptic table with comparative studies and their levels of evidence.

According to evidence-based practice, studies must be evaluated and classified hierarchically according to the level of evidence. This contributes to decision-making in healthcare. In this study, we adopted the proposal of Melnyk and Fineout-Overholt[11], as described below:

I Evidence derived from a systematic review or meta-analysis of all relevant randomized controlled trials or from clinical guidelines based on systematic reviews of randomized controlled trials;

II Evidence derived from at least one clearly delineated randomized controlled clinical trial;

III Evidence obtained from well-designed clinical trials without randomization;

IV Evidence from cohort studies and well--designed case-control;

V Evidence originated from systematic review of descriptive and qualitative studies;

VI Evidence derived from a single descriptive or qualitative study; and,

VII Evidence derived from opinions of authorities and/or reports from expert committees.

RESULTS

Description of the studies

As for the year of publication, of the 16 articles included in the study presented in **Table 1**, there was an increase in publication in 2013, which saw three articles published. Between 2005, 2006 and 2014 no studies related to the theme were identified.

As for the characterization of the studies included in the selection, it should be emphasized that the publications were found in journals of nursing, medicine, hematology, and sciences and public health. All articles were in English and four were national publications.

Regarding the methodological approach, the studies were of quantitative, qualitative, and quantitative and qualitative approach; as for the design, most were cross-sectional, descriptive, and exploratory studies, while there was one case study.

Synopsis of articles included in the study

The selected productions pointed aspects in relation for the care provided to children with SCD and changes to families' routines[7,9,22-23]. Some studies highlighted parents' experiences, specifically the mother, revealing a dual role of mother and caregiver[8,15,23].

The issues involving psychosocial risk factors were another point often found in selected studies[12-13-14-15,21]. These risk factors included emotional changes in the siblings of affected children[14,17,21].

When examining families living with children with SCD, the articles revealed a challenging family operation involving issues such as communication, changed roles, and psychosocial difficulties[7,9,16,20-21].

Table 1 - Description of the studies included in accordance with the identification code of the article / magazine, article title, author, year / language, type of study, and methodological approach. Divinópolis, 2015.

Identification code of the article and magazine	Title	Authors	Year/Language	Type of study	Approach
E01 Rev Paul Pediatr	Quality of life in patients with sickle cell disease.	Menezes ASOP, Len CA,Hilário MOE,Terreri MTRA,Braga JAP	2013 Portuguese/ English	Cross-sectional study	Quantitative
E02 Wis Med J	Parental perception of quality of hospital care for children with sickle cell disease	Kam J,Panepinto JA,Brandow AM,Brousseau DC	2008 English	Cross-sectional study	Quantitative
E03 Haematologica	Quality of life of female caregivers of children with sickle cell disease: a survey	Van Den Tweel XW, Hatzmann J, Ensink E, van der Lee JH, Peters M, Fijnvandraat K, et al.	2008 English	Cross-sectional study	Quantitative
E04 J Pediatr Psychol	Examination of risk and resiliency in a pediatric sickle cell disease population using the psychosocial assessment tool 2.0	Karlson CW,Haynes SL,Smith M,Faith MA,Elkin D,Megason G.	2012 English	Descriptive and exploratory study	Quantitativa
E05 Ciênc Saúde Coletiva	Reproductive decisions and neonatal screening: the prospect of female caregivers of children with sickle cell disease	Guedes C.	2012 Portuguese	Descriptive and exploratory study	Qualitativa
E06 Afr J Psychiatry	Children with sickle cell disease who are experiencing psychosocial problems concurrently with their mothers: a Nigerian study	Ayinmode T.	2011 English	Estudo Transversal e seccional	Quantitative
E07 J Dev Behav Pediatr	Family functioning in the context pediatric chronic conditions	Herzer M, Godiwala N, Hommel KA, Driscoll K, Mitchell M, Crosby LE, et al	2010 English	Cross-sectional study	Quantitative
E08 Issues Compr Pediatr Nurs	Exploring parent-siblings communication in families of children with sickle cell disease	Graff JC, Hankins JS, Hardy BT, Hall HR, Roberts RJ,Neely-Barnes SL.	2010 English	Descriptive and exploratory study	Qualitativa
E09 Pediatr Blood Cancer	Parental information, motivation, and adherence behaviors among children with sickle cell disease	Raphael JL,Butler AM, Rattler TL,Kowalkowski MA, Mueller BU, Giordano TP	2013 English	Estudo transversal	Quantitativo

Gesteira ECR, Bousso RS, Misko MD, Ichikawa CRF, Oliveira PP. Families of children with sickle cell disease: an integrative review. Online braz j nurs [internet] 2016 Jun [cited year month day]; 15 (2):276-290. Available from: http://www.objnursing.uff.br/index.php/nursing/article/view/5289

E10 Pediatr Blood Cancer	The mediating effects of family functioning on psychosocial outcomes in healthy siblings of children with sickle cell disease	Gold JI,Treadwell M,Weissman L, Vichinsky E	2011 English	Descriptive and exploratory study	Quantitativa
E11 J Pediatr Hematol Oncol	Coping and coping assistance among children with sickle cell disease and their parents	Hildenbrand AK,Barakat LP, Alderfer MA, Marsac ML	2015 English	Descriptive and exploratory study	Quantitative and qualitative
E12 Ethn Health	All her children are Born that way: gendered experiences of stigma in families affected by sickle cell disorder in rural Kenya	Marsh VM, Kamuya DM, Molyneux SS	2013 English	Descriptive and exploratory study	Qualitative
E13 East Afr Med J	Psychosocial impacto f sickle cell disease on mothers of affected children seen at University of Ilorin Teaching Hospital, Ilorin, Nigeria	Tunde-Ayinmode MF	2007 English	Cross-sectional study	Quantitative
E14 Rev Bras Hematol Hemoter	The daily life of families of children and adolescents with sickle cell anemia	Guimaraes TMR, Miranda WL, Tavares MMF	**2009 Portuguese**	**Descriptive and exploratory study**	**Qualitative**
E15 Rev Eletr Enf	**The daily life of a family experiencing a chronic condition due to sickle cell anemia**	**Silva AH, Bellato R, Araujo LFS**	**2009 Portuguese**	**Case study**	**Qualitative**
E16 Clin Pediatr (Phila).	**Parent perspectives on pain management, coping, and family functioning in pediatric sickle cell disease**	**Mitchell MJ, Lemanek K, Palermo TM, Crosby LE, Nichols A, Powers SW**	**2007 English**	**Descriptive and Exploratory Study**	**Quantitative and qualitative**

Source: Authors.

There were also studies on children's and parents' (caregivers) quality of life[9,13,22,25].

Another aspect raised in the articles was related to the origin of the disease, which is related to issues of ethnicity, gender, and stigma since the disease originated in the population of African descent[12.19].

The need to improve social support and support networks for children and their families was also highlighted in the studies[7-8-9,18,20,21,24,25].

A comparative synopsis of the studies and their levels of evidence are shown in **Table 2**.

DISCUSSION

From the analysis of the content of selected publications, four thematic categories

Table 2 - Summary of articles according to the reference/code study, participants objectives and results. Divinópolis, 2015.

Code and reference of the study	Population/ Sample	Objectives	Results	Evidence Level
E01(22)	100 patients with sickle cell disease (SCD) and their parents	To evaluate the quality of life related to the health of children and adolescents with SCD	The SCD compromises the quality of life of children, adolescents and their families	V
E02(12)	Parents of 112 children hospitalized with SCD.	Assess how parents perceive the quality of care in the hospital	Parents of children with SCD realize that there is a low quality in terms of hospital care for their children related to the fact that the disease is originally afrodescendant	V
E03(13)	54 caregivers of children with SCD.	To evaluate the quality of life of caregivers of children with SCD	Caregivers of children with SCD have significantly lower quality of life at all scales of the questionnaire compared with the control group, including depressive states, daily activities, and vitality.	II
E04(14)	219 caregivers of children with SCD.	Assess the risk of psychological distress in families of children with SCD	The emotional problems of the sickle cell patients affect the brothers and family members and are psychosocial risk factors for the whole family.	II
E05(23)	50 women caregivers of children with SCD	To analyze the perception of reproductive risk in women and caregivers of children with SCD.	Women want to have other children even at the risk; women do not want to have children for fear of another child suffering from SCD.	V
E06(15)	250 mothers (100 mothers with children suffering from SCD)	Identify children with SCD who experience psychosocial problems with their mothers	There was a correlation between children with probable tendency to psychological problems and mothers.	II
E07(16)	301 parents of children with disorders. Chronicles, and 44 suffer from SCD.	Describe and compare the overall family functioning of children with five chronic diseases (including the SCD)	The operation is unhealthy and is related to risk factors such as: child age, the small number of children living in the home, and low income.	II
E08(17)	52 parents of children with SCD	Explore how the communication between parents and siblings of children with SCD occurs and to identify factors that influence it.	Communication is effective, they show interest, awareness and responsibility in caring for siblings with SCD.	V

Gesteira ECR, Bousso RS, Misko MD, Ichikawa CRF, Oliveira PP. Families of children with sickle cell disease: an integrative review. Online braz j nurs [internet] 2016 Jun [cited year month day]; 15 (2):276-290. Available from: http://www.objnursing.uff.br/index.php/nursing/article/view/5289

E09(18)	150 parents of children with SCD	Examine the correlation of psychosocial variables of parents caring for their children with SCD	The correlations of psychosocial variables: young age of the child, use of health services, parental age, and marital status influence care.	II
E10(7)	65 families of children with SCD.	Examine the role of family functioning on the psychosocial functioning of the healthy siblings of children with SCD.	The study showed that behavioral and psychological difficulties impair family functioning.	V
E11(9)	Parents of 15 children with SCD.	Identify the stressors of SCD and how parents face these factors.	Stressors are medical complications, side effects of treatment, interruption of daily routines and activities, emotional reactions, communication issues, social challenges, and worries about the future.	II
E12(19)	13 family members from rural Kenya with SCD	Explore the families of experiences with children affected by sickle cell anemia and how these experiences influence relationships in the family	The study indicates that within families, mothers suffer stigmatization by being seen as bearers of SCD for their children.	V
E13(20)	Study involving 100 mothers of children with SCD	Assess the psychosocial impact of SCD on mothers.	28% of mothers of children with SCD were identified as probable cases of psychological problems.	V
E14(8)	Interviews with 10 families with at least one child with sickle cell anemia (SCA)	Analyze the daily lives of families of children with SCA	SCA affects the entire family, and the burden of care falls on the mother which changes the family dynamics.	II
E15(24)	Case study of a family with a child suffering from SCA	Understanding the daily life of a family experiencing a chronic condition due to sickle cell anemia	The family is affected in their daily lives by the need for continuity of care.	V
E16(21)	53 parents of children with SCD	Check the relationship between patient and family coping and healthcare of children with SCD	The positive coping patient was related to positive family functioning and lower use of health services.	VI

Source: Authors, 2015.

emerged: the impact of SCD on the family; the psychosocial effects of the disease on family dynamics; the quality of life of the child and its family; and, social support and support networks.

SCD's impact on the family

Each family is a unit and we need to pay attention to interactions among family members, not just the individual, when in contact with it. Therefore, in the management of chronic disea-

se, assessment and care should include a focus on the family, not just the patient[15].

One study confirmed that SCD changes family routine, modifying not only the child's life, but also that of their healthy siblings who experience interruptions in their routines because of the demands of the affected sibling's disease. Consequently, there is an imbalance in the stress and psychosocial functioning of family members[7]. These changes in the household are triggered by a new lifestyle attached to the care related to specific drug use[13,23], constant hospitalizations during times of crisis[22], the search for a proper diet[23], restrictions on physical activity[9,23], and other precautions that are necessary to control the morbidity[23] related to SCD.

These modifications imposed by the management of the disease overload parents, who not only have to take care of the child affected by the disease, but also need to balance the care needed by their other healthy children. This can lead to conflicts in a family's adjustment[14,17,21].

In this sense, families and children experience various feelings, including suffering caused by the physical and psychosocial burden, which can affect all members, but especially the primary caregiver[9].

By studying the daily lives of families, researchers stressed that mothers are primary caregivers[8] who assume the dual role of mother and caregiver[8,15,23] since they are more involved in the daily care of their children[15].

Another study[23] showed that the experience of care aimed at children with SCD determines the reproductive decisions of mothers—some do not wish to have more children for fear of having a second child with the disease. Having other children would also entail more expense, requiring a greater increase in family income[13].

Another study[8] highlighted that many mothers leave work to devote themselves exclusively to their sick child, often looking for informal jobs that can be undertaken in their own homes to ensure they can give close and continuous attention to their child; therefore, a reduction in family finances[15,23] can create risks of physical, social, and emotional distress leading to marital discord[15].

Caring for children with SCD involves developing daily coping mechanisms[8]. Families seek the best way to face and adapt to a life marked by worsening episodes of the disease[12], suffering, fear of the future, absence of employment, and events that trigger psychosocial risks[8,12-13].

Psychosocial effects on family dynamics

The psychosocial effects of caring for a child with SCD can be generated by guilt at having gestated a sick child[9,19]. Moreover, the risk of conceiving another child with SCD may have a negative effect on the mood of caregiving mothers[7,22].

Many families of children with SCD face psychosocial challenges related to frequent hospitalizations and restrictions on social activities that trigger impairment to quality of life and psychological symptoms[9].

The stress of disease management has repercussions on family dynamics, which may lead to a risk of depression in parents[22], as highlighted in an article that states that half of the caregivers of children with SCD are at risk of depression[13].

Studies of American families from Africa stressed that financial difficulties[7,17,19-20,22], caregivers' low level of education[13,19], and difficult family functioning[7,15-16,] are crucial risk factors in increases in disease symptoms and families' psychological distress[14].

Gesteira ECR, Bousso RS, Misko MD, Ichikawa CRF, Oliveira PP. Families of children with sickle cell disease: an integrative review. Online braz j nurs [internet] 2016 Jun [cited year month day]; 15 (2):276-290. Available from: http://www.objnursing.uff.br/index.php/nursing/article/view/5289

However, several articles[15,20] claimed that the management of SCD requires special care for children even without it being in crisis. Combined with the intermittent experiences of crises, hospital admissions, and continuous transfusions, it is remarkable that stress poses a risk for the psychosocial dysfunction of caregivers and other family members.

As with any chronic illness, stress and suffering can lead to interruptions in intra-family relations[7]. Families seek a new direction to work towards more organized family functioning regarding the psychosocial effects that arise.

In this sense, nursing is placed in a privileged position. As an area of care for excellence, we can contribute with guidelines that may help families in adapting to their new situation[12].

The functioning of families with children with SCD reduces families' quality of time together due to the amount of hospitalization periods and the need for medical care, undermining emotional involvement among family members[16,21]. Families can benefit from strategies to enhance communication, time management, and conflict resolution—that is, family functioning. They can do so through improving and negotiating their roles before and during health crises in order to promote better management of the disease[16].

To deal with family dysfunction, authors have developed research in this field and have suggested that greater attention should be paid to the areas of communication[16-17], roles[14,21], and emotional involvement between family members[16]—including sick children's siblings, who, when included in two-way communication with their parents[17], have demonstrated knowledge in terms of the care of their affected sibling.

Thus, the involvement of all family members in sick children's care alleviates the caregiver's burden, facilitates the division of tasks and roles, organizes family functioning, and reduces symptoms such as the risk of depression[13,22], stress, and suffering in children[7,8,12-13-14-15] and their families.

Quality of life for children and their families

International[13.16] and national[22] publications that focus on children's and families' quality of life have lead to reflections on this issue due to restrictions in emotional, social, physical, and family aspects surrounding them in their daily lives.

The theme has been investigated since SCD is a chronic disease that leads to limitations[8-9,13,24], frustration, despair, and loss[8-9,22,24]. Children fail to attend school when they are in crisis or need hospital care[8-9,12-13,16,21-22] and the physical and psychosocial burdens lead to psychological illnesses that impair the quality of life of these people[8-9,13,18,21-22].

Over the past 20 years we have developed several tools to evaluate children's quality of life. One of them is the Pediatric Quality of Life Inventory (Peds QL), version 4.0, which evaluates the quality of life of healthy children and those with chronic diseases. This was recently validated in the Brazilian context[22].

Studying the quality of life of children with SCD is necessary since a number of factors, such as pain, affect the quality of life of these children as the disease is associated with loss of recreational and social functioning, as well as school attendance and leisure activities, which impairs daily life[7,21].

As important as evaluating the quality of life in children with SCD is the need to evaluate their caregiver parents who also suffer the difficulties of the daily management of the

disease[22], as well as dealing with the burden of having transmitted a disease to their child that involves social and ethnic issues[9,13,19]. A survey showed that some women are stigmatized by their families because they are seen as carriers of the disease[19]. This increases the risk of a poor quality of life for these women[14].

Another study identified that parents perceive a lower quality in the care provided to their children in the hospital environment and believe that this service is related to ethnic and racial issues[12]. This corroborates the findings of the previous study, which highlights that African descent origin leads to stigmatization[19] and thus reduces the quality of life of caregivers and children with SCD[14].

Among the determinants that contribute to poor quality of life are socioeconomic aspects[22] due to the fact that most caregivers fail to work or ask for early leave in order to care for their sick child[8,24], which usually results in a caregiver overload[22], decreasing their daily quality of life.

It is worth emphasizing the need for further studies in the Brazilian context in order to analyze the theme *Quality of life of children with SCD* and the socioeconomic and cultural variables in order to highlight gaps and improve comprehensive care for these patients and their families[22].

Social support and support networks for families of children with SCD

Considering healthcare and the need for family assistance, international studies claim that social support is crucial for adherence to treatment[7,9,18].

After confirming the diagnosis, the family's motivation to undergo therapy for the child suffering from SCD is facilitated by the confrontation before the stressful events that permeate the care routine of the child, such as recurrent hospitalizations, examinations, and consultations[18]. Therefore, social support is the foundation for parents and children to manage SCD with more security[8]. Limited social support discourages families, who, helplessly, still struggle to make decisions regarding their child's treatment regimen[14,24].

Thus, extended family, friends, neighbors, and health professionals, as pillars of social support[18], favor the development of parental skills in order to face their modified routine due to their child's disease[8,18].

Families' behavior toward the health and disease process is influenced by different types of support: emotional, social, and medical. More social support corresponds to a greater involvement and family awareness in terms of the care provided to the sick child. In case there are behavioral and psychological difficulties regarding the daily management of children with SCD, there is also the misfit in family dynamics and some families may stop exercising the routine of care[7,15,16], increasing the risk of disease complications[7].

To achieve resilience, a study pointed out that relaxation among family members is an effective strategy for social support and support networks because when grandparents come to assume the management of care[14], there is a decreased risk of psychosocial problems within the family[8,12-13].

In the context of the coping process of families, social support includes strategies for decision making, acceptance, and management of the disease[14,24]. These strategies are achieved in a social environment, which includes as breadwinners religion and spirituality[8,14,24].

In this sense, the participation of families of children with SCD in social organizations and associations for SCD patients[8,20] streng-

Gesteira ECR, Bousso RS, Misko MD, Ichikawa CRF, Oliveira PP. Families of children with sickle cell disease: an integrative review. Online braz j nurs [internet] 2016 Jun [cited year month day]; 15 (2):276-290. Available from: http://www.objnursing.uff.br/index.php/nursing/article/view/5289

thens families, since spaces for sharing experiences allow for the sharing of problems that are common to families' routines[8,24].

We should not ignore the interconnected services that support families of children with SCD, such as clinics, hospitals, hematology centers, and family health strategies[24]. It is legitimate for certain families to feel satisfied with some services and thus create emotional bonds, while in other cases a family's connection with a service unit can be low[24], thus the family does not feel at ease and holds little confidence in the service, preventing the expression of feelings. This complicates families' adaptation in the management of SCD[8].

In the context of services as sources of support for these families, it should also be emphasized that, due to the chronic nature of SCD, it is essential to achieve shared monitoring with the Primary Health Care (PHC) support network[24] through an effective system of reference and counter with the establishment of a care plan that provides the most effective support between family and service. This way it would facilitate the achievement of comprehensive care[24], that is, interconnected assistance between the APS units and Hematology Centers, given that one of the principles of the *Sistema Único de Saúde* (SUS) (Unified Health System) is the entirety described in the line of care to patients with SCD in the manual created by the Ministry of Health[24].

Within the principle of comprehensiveness, monitoring patients and families should be offered by the primary care network and not only for hematology services, since one of the principles of the SUS is decentralization, indicating that its members should be served close to their homes[24].

This principle is faced with the limited participation of health professionals in the care of children with SCD and their families[24]; that is, the permanent education of professionals is necessary so that the goal of comprehensive care provided to the child and its family is effectively achieved[8,24].

However, with a healthcare system that is still fragmented, families remain the leading providers of care for children with SCD[8,22], trying to meet the challenges involved in the achievement of comprehensive care[24] and fighting for the quality of life of children living with the disease[8].

CONCLUSION

One of the limits of this research is that is was restricted to low levels of national research on the subject of families of children with SCD, especially studies related to families' daily management of the disease.

It is also true that few studies in the nursing field were found and, as SCD is one of the most common hereditary diseases in Brazil, it requires continuous care from a multidisciplinary team, which includes nurses.

Before the impact of SCD in families, we emphasize the importance of a study that corroborates the reflections of health professionals who assist children with SCD, revealing the importance of the family in the context of care and their real needs in terms of managing the disease.

In the analyzed articles we can see the need for studies that address the evaluation of families in terms of care experience, with research showing the use of assessment tools with families to provide support to the knowledge of the internal structure, support networks, and support of families when caring for a child with SCD. Moreover, nursing should propose interventions tailored to each

family through the steps of problem raising, care planning, and the implementation and evaluation of interventions that contribute to an effective systematization of primary, secondary, and tertiary nursing care to the carrier of SCD and his or her family.

It has been found that scientific research on the families of children with SCD in Brazil is still insufficient with regards to comprehensive care as there are still difficulties in integrating assistance in the sphere of primary care with hematology centers where children with SCD undergo consultations, tests, and transfusions. There is little communication between the services to ensure a continuity of care, which fragments care.

For family management, national and international studies have shown that events occurring around the disease and care require coping strategies and family adaptation.

In the literature, there is a wealth of studies that explore the psychosocial risk factors for child and family, reviews of quality of life (mainly indicating the mother as the primary caregiver), and surveys of socioeconomic and cultural factors that involve populations of African origin.

We should also note that research on family functioning and family responses to the disease and the care of SCD need to be further studied by health professionals, specifically nursing, in terms of their approach to families in care. As mentioned in the limitations of this study, it is necessary to establish personalized interventions according to the needs of each family based on studies that focus on the functioning of family dynamics.

It is believed that a better understanding of the demands and wishes of families and the identification of elements that hinder the daily management of the disease, such as psychosocial risk factors listed by the authors of this integrative review, can stimulate and support multidisciplinary teams in building a continuum of care to families, which will facilitate achievement in terms of the health of children with SCD in its entirety and a quality of life similar to that of other children.

REFERENCES

1. Jordan L, Swerdlow P, Coates TD. Systematic review of transition from adolescent to adult care in patients with sickle cell disease. J Pediatr Hematol Oncol. 2013; 35(3):165-9.
2. Vigilante JA, DiGeorge NW. Sickle cell trait and diving: review and recommendations. Undersea Hyperb Med. 2014; 41(3):223-8.
3. Rodrigues DOW, Ferreira MCB, Campos SEM, Pereira PM, Oliveira CM, Teixeira MTB. História da triagem neonatal para doença falciforme no Brasil. Rev Med Minas Gerais. 2012; 22(1):66-72.
4. Silva-Pinto AC, Angulo IL, Brunetta DM, Neves FI, Bassi SC, Santis GC, et al. Clinical and hematological effects of hydroxyurea therapy in sickle cell patients: a single-center experience in Brazil. São Paulo Med J. 2013; 131(4):238-43.
5. Kanter J, Kruse-Jarres R. Management of sickle cell disease from childhood through adulthood. Blood Rev. 2013; 27(6):279-87.
6. Gold JI, Treadwell M, Weissman L, Vichinsky E. The mediating effects of family functioning on psychosocial outcomes in healthy siblings of children with sickle cell disease. Pediatr Blood Cancer. 2011; 57(1):1055-61.[included in the review].
7. Guimaraes TMR, Miranda WL, Tavares MMF. The day-to-day life of families with children and adolescents with sickle cell anemia. Rev Bras Hematol Hemoter.2009;31(1):9-14. [included in the review].
8. Hildenbrand AK, Barakat LP, Alderfer MA, Marsac ML. Coping and coping assistance among children with sickle cell disease and their parents. J Pediatr Hematol Oncol. 2015; 37(1):25-34. [included in the review]

Gesteira ECR, Bousso RS, Misko MD, Ichikawa CRF, Oliveira PP. Families of children with sickle cell disease: an integrative review. Online braz j nurs [internet] 2016 Jun [cited year month day]; 15 (2):276-290. Available from: http://www.objnursing.uff.br/index.php/nursing/article/view/5289

9. Whittemore R, Knafl K. The integrative review: updated methodology. J Adv Nurs. 2005;52(5):546-53.
10. Melnyk BM, Fineout-Overholt E. Making the case for evidence-based practice. In: Melnyk BM, Fineout-Overholt E. Evidence-based practice in nursing & healthcare. A guide to best practice. Philadelphia: Lippincot Williams & Wilkins; 2005. p. 3-24.
11. Kam J,Panepinto JA,Brandow AM,Brousseau DC. Parental perception of quality of hospital care for children with sickle cell disease. Wis Med J. 2008;107(3):131-35. [included in the review]
12. van den Tweel XW, Hatzmann J, Ensink E, van der Lee JH, Peters M, Fijnvandraat K, et al. Quality of life of female caregivers of children with sickle cell disease: a survey. Haematologica. 2008;93(4):588-93. [included in the review]
13.Karlson CW, Haynes SL, Smith M, Faith MA, Elkin D, Megason G. Examination of risk and resiliency in a pediatric sickle cell disease population using the psychosocial assessment tool 2.0. J Pediatr Psychol. 2012; 37(9):1031-40. [included in the review]
14. Ayinmode T. Children with sickle cell disease who are experiencing psychosocial problems concurrently with their mothers: a Nigerian study. Afr J Psychiatry (Johannesbg). 2011; 14(1):392-401. [included in the review]
15.Herzer M, Godiwala N, Hommel KA, Driscoll K, Mitchell M, Crosby LE, et al. Family functioning in the contexto f pediatric chronic conditions. J Dev Behav Pediatr. 2010; 31(1):1-14. [included in the review]
16.Graff JC, Hankins JS, Hardy BT, Hall HR, Roberts RJ, Neely-Barnes SL. Exploring parent-siblings communication in families of children with sickle cell disease. Issues Compr Pediatr Nurs. 2010; 33(2):101-23. [included in the review]
17.Raphael JL, Butler AM, Rattler TL, Kowalkowski MA, Mueller BU, Giordano TP. Parental information, motivation, and adherence behaviors among children with sickle cell disease. Pediatr Blood Cancer. 2013; 60(7):1204-10. [included in the review]
18. Marsh VM, Kamuya DM, Molyneux SS. "All her children are born that way": gendered experiences of stigma in families affected by sickle cell disorder in rural Kenya. Ethn Health. 2011; 16(4-5):343-59. [included in the review]
19. Tunde-Ayinmode MF. Psychosocial impact of sickle cell disease on mothers of affected children seen at University of Ilorin Teaching Hospital, Ilorin, Nigeria. East Afr Med J. 2007; 84(9):410-19. [included in the review]
20. Mitchell MJ, Lemanek K, Palermo TM, Crosby LE, Nichols A, Powers SW. Parent perspectives on pain management, coping, and family functioning in pediatric sickle cell disease. Clin Pediatr (Phila). 2007;46(4):311-19. [included in the review]
21. Menezes ASOP, Len CA, Hilário MOE, Terreri MTRA, Braga JAP. Quality of life in patients with sickle cell disease. Rev Paul Pediatr. 2013;31(1):24-9. [included in the review]
22. Guedes C. Decisões reprodutivas e triagem neonatal: a perspectiva de mulheres cuidadoras de crianças com doença falciforme. Ciênc Saúde Coletiva. 2012; 17(9): 2367-76. [included in the review]
23. Silva AH, Bellato R, Araújo LFS. Cotidiano da família que experiência a condição crônica por anemia falciforme. Rev Eletr Enf. [internet]. 2013 Apr-Jun [Cited 2015 May 09]; 15(2):437-46. Available from: http://dx.doi.org/10.5216/ree.v15i2.17687. [included in the review]
24. Rodrigues CCM, Araújo IEM, Melo LL. A família da criança com doença falciforme e a equipe enfermagem: revisão crítica. Rev Bras Hematol Hemoter. 2010;32(3):257-64. [included in the review]
25. Wright LM, Leahey M. Enfermeiras e Famílias: um guia para avaliação e intervenção na família. 5. ed. São Paulo: Roca, 2012.
26. Carvalho AS, Depianti JRB, Silva LF, Aguiar RCB, Monteiro ACM. Reactions of family members of children diagnosed with cancer: a descriptive study. Online braz j nurs [Internet]. 2014 Sep [Cited 2015 Mar 20]; 13(3):282-91. Available from: http://www.objnursing.uff.br/index.php/nursing/article/view/4356

27. Amaral JL, Almeida NA, Santos PS, Oliveira PP, Lanza FM. Perfil sociodemográfico, econômico e de saúde de adultos com doença falciforme. Rev Rene. 2015; 16(3):296-305

All authors participated in the phases of this publication in one or more of the following steps, in According to the recommendations of the International Committee of Medical Journal Editors (ICMJE, 2013): (a) substantial involvement in the planning or preparation of the manuscript or in the collection, analysis or interpretation of data; (b) preparation of the manuscript or conducting critical revision of intellectual content; (c) approval of the versión submitted of this manuscript. All authors declare for the appropriate purposes that the responsibilities related to all aspects of the manuscript submitted to OBJN are yours. They ensure that issues related to the accuracy or integrity of any part of the article were properly investigated and resolved. Therefore, they exempt the OBJN of any participation whatsoever in any imbroglios concerning the content under consideration. All authors declare that they have no conflict of interest of financial or personal nature concerning this manuscript which may influence the writing and/or interpretation of the findings. This statement has been digitally signed by all authors as recommended by the ICMJE, whose model is available in http://www.objnursing.uff.br/normas/DUDE_eng_13-06-2013.pdf

Received: 08/10/2015
Revised: 04/27/2016
Approved: 04/27/2016

Gesteira ECR, Bousso RS, Misko MD, Ichikawa CRF, Oliveira PP. Families of children with sickle cell disease: an integrative review. Online braz j nurs [internet] 2016 Jun [cited year month day]; 15 (2):276-290. Available from: http://www.objnursing.uff.br/index.php/nursing/article/view/5289