

The participation of cohabitants with sickle cell disease in health care: a bibliographic study

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Abstract *This paper aims to analyze the published scientific production about the participation of the sickle cell disease (SCD) subjects and their relatives, and the autonomy and social aspects of these individuals. A qualitative bibliographic search with the Portuguese-equivalent keywords “sickle cell disease” and “participation” was used. As a result, the following themes appeared: (1) Experience of illness, highlighting coexistence and ethnic-racial issues; (2) Participation in research and the perspective of health professionals on SCD; and (3) Autonomy of cohabitants and decision-making. We can conclude that the promotion of the participation of these patients in the studies, either instrumentally, or to contextualize the results better, or – still – to enrich the authors’ conclusions, can intentionally or unintentionally contribute to the greater visibility of the problem that involves being a SCD cohabitant for the subjects and their relatives. The duty to analyze intersectionally the entire context of the patient and his family context is also highlighted.*

Key words *Sickle cell disease, Participation, Literature review, Qualitative research*

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Introduction

Sickle cell disease (SCD) is a group of hemoglobinopathies characterized by structural changes in blood cells that produce an abnormal hemoglobin HbS, and the 'S' derives from "sickle", due to the shape of the hemolytic cells of the diseased patients^{1,2}.

In academic literature, the terms designating the disease vary widely. Terms like "sickle cell anemia", "sickle cell syndrome", "sickle cell disease" and "drepanocytosis" can be found. According to Silva³, the term sickle cell disease has been accepted for expressing sickle cell anemia (a condition with symptoms) and other disorders caused by the same mutation.

SCD is defined as a recessive genetic disease. Patients with sickle cell anemia inherit the HbS from their parents, considered simple heterozygotes, that is, patients with the sickle cell trait (without expressed symptoms)⁴.

The World Health Organization reports that about 5% of the world population is affected by hematological diseases such as SCD. In Brazil, 2,000,000 people are estimated with the HbS gene, such as the sickle cell trait, and 8,000 with the disease's homozygosity and its varied clinical implications^{5,6}.

In Brazil, SCD diagnosis can occur from the National Neonatal Screening Program (known as "Baby Foot Test"), which, while not widely implemented, can be crucial for treatment and referral to specialized care teams⁷. Late diagnoses, on the other hand, are made in the face of some problems, causing significant damage to patients, due to the complex symptoms, and SCD can be confused with other diseases such as epilepsy and rheumatoid arthritis⁸.

Far beyond the clinical implications and painful crises⁹, SCD causes striking conflicts and social impacts for patients and their relatives, and fall into the category of chronic illness and require specific diagnosis, consent to treatment, self-care measures and a new lifestyle¹⁰.

The repercussion of the chronic illness generates feelings of insecurity, fear of loss, and frustrations in everyone around the patient¹¹, especially in cases directly affecting children. Anxiety, aggression, depression, and social limitations can be overcome¹² with cooperation in the family environment.

Due to the high SCD incidence in the black population, there may be intense discrimination in an actively racist society and a health system marked by institutional racism¹³. Silva¹⁴ discusses

racial clashes with their problems about the racialization and stigmatization of patients with SCD as belonging to black bodies in the North American context, which both implied racial discrimination, and served as a flag to claim measures from the state that favored blacks.

If we use Goffman's perspective¹⁵, the SCD subjects (because they live with chronic illness) can be stigmatized and placed in a "non-place" by those socially considered "normal", determining invisibility and marginalization of patients with SCD, exempting them thus from a leading posture in therapeutic decisions.

Goffman¹⁵ uses stigma more broadly from the original meaning coined by the Greeks, which was linked to body marks. He said this expression refers to a derogatory attribute, without considering a relationship of attributes in itself, but instead a relationship of language. In this sense, while some cases have no visible physical traumas, people with SCD are stigmatized as social norm deviants. The relativity of Goffman's concept¹⁵ where what can be considered stigma for some, can fit into a standard of normality for others, given that "the normal and the stigmatized are not individuals per se, but rather perspectives, viewpoints, interpretations, and interactions"¹⁶(p.51) is emphasized.

The stigmatized position of those living with SCD can be an expression of the lack of information in their means of living. According to some health professionals, for example, opiophobia – fear of using opioids and their addictive potential – can lead to difficulties in the relationship and access to health services by SCD patients¹⁷.

It is necessary to comment on the intersectionality of these positions: race, social class, and chronic condition. Crenshaw¹⁸ affirms that the concept of intersectionality comes to the union of oppression without established hierarchies, ensuring the inseparability of positions and the need to fight for all the rights inherent to the conditions imposed.

Alves¹⁹ affirms that we have the Talcott Parsons' theory, where the illness was also seen as a deviation, and it was feasible to predict behavior for the patient, with rights and duties, both for patients and for medical conduct in the face of illness. Today, we still find remnants of this theory, especially with the substantial medical authority in decision-making, without considering the autonomy of patients in their treatment process.

This attitude in the field of health professionals disregards any biographical situation and

knowledge stock of patients at an alarming level. In the light of Schutz²⁰, these concepts are related to a catalog of experiences lived by patients who can infer interpretations and actions aimed at a particular aspect of their lives.

The substantial social implications aimed to understand the cultural and societal phenomena linked to SCD call for a broad discussion. In this sense, it is necessary to focus on the participation in SCD care of the subjects of this disease and their relatives (associated or not in a group) so that patient care and integration actions in the decision-making process can be considered or reframed.

The concept of participation discussed here refers not only to the participation of cohabitants in advisory councils in order to guarantee rights, as advocated by the principles of the SUS, but mainly to the participation of patients who, in the process of sensitizing and overcoming imposed stigmas, become the leading figures of their rights and active agents in their treatment.

From this perspective, we aim to analyze in this work the published scientific production about the participation of SCD individuals and their relatives in the treatment, and about the autonomy and social aspects related to these subjects.

Methods

This study consists of bibliographic research, here understood generically as that which uses published secondary sources to analyze the contributions of several authors on a subject²¹. The preferred source here is the scientific paper because it allows higher access in comparison with other types of bibliographic sources.

The Virtual Health Library (BVS - <http://brasil.bvs.br/>) was employed because it is a metasearch engine that can systematize data from other databases such as SciELO, PubMed and Lilacs, relevant databases for scientific knowledge in Brazil and other countries. The high visibility and reliability of this academic metasearch engine, which is part of an initiative of the Brazilian Ministry of Health, the World Health Organization, and the Pan American Health Organization, stands out.

The term used in the academic literature for patients with sickle cell disease is quite diverse and mostly does not cover their family context, and represents only individuals directly affected by this disease. This review proposes the Portu-

guese-equivalent term “living with the SCD” because it understands that the disease addressed here comprises much more than a single individual, but instead all of the means of living in his/her support networks with high intensity.

Following a model of the movement of people with disabilities and the discourse of people living with HIV/AIDS, the term “carrier” was not employed in this paper because the verb “carry” can be removed, not contextualized to the SCD²². Moreover, the term “carriers” refers to the geneticist logic, which reinforces the genetic inheritance imposed by the SCD, reflecting not only the symptomatological disease but also the sickle cell trait, avoiding any questioning about the accountability or guilt of the genetic discourse.

Other terms were used, but they must be discussed. The term patient was used in this review comprehensively and without reinforcing the biomedical and clinical logic, as this is socio-anthropological research. “SCD subjects” was used as a synonym for “coexisting with the SCD” because they believe that people engaged in the theme do not necessarily need to be directly affected by the disease.

The subject descriptors used for this analysis were undoubtedly aligned with the central core of the objective of this paper (SCD and participation). It is noteworthy that, considering the representation of patient movements and mobilization for the benefit of the SCD, the term “patient association”, related to the theme proposed here, was excluded as a descriptor because it linked as results only the association of symptoms, medications or treatments referring to sickle cell disease.

The term “*participação*” was determined to define public participation or community action under the BVS’ subject descriptors in Portuguese. As synonyms of the term, the database points to “*Participação Comunitária*”, “*Participação Popular*”, “*Mobilização*” among others. The term “*Doença Falciforme*” appears as equivalent to “*Anemia Falciforme*”, “*Anemia de Células Falciformes*”, “*Doença das Células Falciformes*”, among others.

The search carried out on December 29, 2017, under the Portuguese terms “*doença falciforme*” and “*participação*”, resulted in 34 references. The inclusion criteria in this review were: (1) classification as a scientific paper (excluding theses and editorials), (2) year of publication between 2000 and 2017, and mainly (3) be defined as qualitative research, thus excluding clinical, epidemiological and quantitative studies in general, which left out 20 papers.

A new search was carried out on March 1, 2018, to verify whether new papers with the profile of this review were included in the database, which was not observed. Works published before 2000 were excluded from this research for better analysis and sample restriction. This time constraint was due to the limited production available on online platforms. In the research, only one paper was identified before 2000, of 1973, which addresses laws and policies based on genetic technologies of that time, which does not contribute significantly to the theme proposed here.

Concerning the analysis of the qualitative collection found, we adopted the thematic analysis technique adapted by Gomes²³, based on the principles of Bardin²⁵. The central concept of this analysis is the theme that “comprises a bundle of relationships and can be graphically presented through a word, sentence or summary”²³(p.86). The following analytical trajectory was followed as a basis for this author: (a) identification of the central ideas of the excerpts transcribed from all papers, (b) classification of the meanings underlying the ideas in themes that summarize the production of knowledge about the studied subject, and (c) elaboration of interpretative syntheses of each theme, establishing a dialogue with the themes identified in the analysis of the papers with other works – pointed out in the introduction of this work – so that the discussion on the subject could be expanded.

Results and discussion

With the results of the search, Table 1 presents the list of papers analyzed, followed by the year of publication, country of origin, and prominent themes. Regarding the country of origin, a low frequency of studies was identified in the southern hemisphere, which can be explained not necessarily by the low production of these countries, but perhaps because this production does not always meet BVS' indexing criteria. Two studies^{23,24} from Kenya stand out for addressing a non-Eurocentric or Americanized scenario in dealing with genetic information issues.

Noteworthy is the fact that most of the works analyzed here aimed at those living with SCD, looking at interviews and consultations on popular participation, therapeutic options, among other topics raised in this study. Only three papers²⁵⁻²⁷ used health professionals involved in the SCD theme as their research object.

We emphasize here that some papers addressed more than one theme included in this work, although the year-to-year variation has become quite homogeneous, and 2010 had five publications under the analyzed descriptors.

The following themes emerged from the content analysis of the papers: (1) Experience of illness, related to living with the SCD and ethnic-racial issues; (2) Participation in research and the perspective of health professionals on SCD and (3) Autonomy of cohabitants and decision-making. These themes – which are not necessarily mutually exclusive – are directly or indirectly related to the participation of those living with the SCD.

Experience of illness

This theme is characterized by the patient's experience and perspective in the face of illness due to SCD.

Silva et al.²⁸ address the family's crucial role (as a “care-producing unit” in coping with chronicity) and the networks that support the family involved in the research. Formal and informal care institutions and their contribution to maintaining the daily lives of cohabitants, such as extended family (grandparents, uncles, among others), patient associations, and religious support, are highlighted in the research with the representation on an ecomap. These authors emphasize the role of patient associations as a space for sharing experiences and strengthening bonds and affection among peers.

Doulton²⁹ describes the program for the transition of patients with SCD from childhood to adulthood, pointing out the significant advances in medicine, where the life expectancy of patients with SCD has increased significantly, from 14 to 50 years²⁹. Thus, it becomes necessary to adapt services and care to this new reality. The author proposes health education centered on self-care and emphasizes the need for an analysis of the experiences of cohabitants, not reducing the disease, but observing cultural, socioeconomic aspects, and the entire family context. They also consider the subjects' attempts to adapt and fit into the socially imposed functionality.

Willien et al.³⁰ present a review aimed at SCD patients, addressing uncertainties and possible complications experienced by illness and its potential complications such as the possibility of infections, crises under changes in altitude, and routine vaccinations. The authors and Doulton²⁹ emphasize the need to create measures that ac-

Chart 1. Characterization of the analyzed papers.

Study	Year	Country/Origin	Theme
Noke et al. ²⁵	2016	United Kingdom	Health professionals
Patterson et al. ⁴¹	2015	United States	Participation in research
Marsh et al. ²³	2013	United Kingdom	Health professionals
Silva et al. ²⁸	2013	Brazil	Illness experience
Lebensburger et al. ⁴²	2013	United States	Participation in research
Jones e Broome ⁵⁰	2001	United States	Participation in research/Autonomy of cohabitants and decision-making
Daulton ²⁹	2010	United States	Illness experience
Reed ³⁹	2011	United Kingdom	Illness experience
Marsh et al. ²⁴	2013	Kenya / United States	Health professionals
Fry ³⁸	2005	Brazil	Illness experience
Tsianakas et al. ²⁶	2010	United Kingdom	Health professionals
Benjamin ⁴³	2011	United States	Participation in research
Liem et al. ⁴⁴	2010	United States	Participation in research
Lattimer ⁴⁰	2010	United States	Illness experience/Autonomy of cohabitants and decision-making
Bakshi et al. ²⁷	2017	United States	Health professionals
Hankins ⁴⁵	2010	United States	Participation in research
Hassel et al. ⁴⁶	2008	United States	Participation in research/Autonomy of cohabitants and decision-making
Willen et al. ³⁰	2014	United States	Illness experience
Hankins et al. ⁴⁸	2007	United States	Autonomy of cohabitants and decision-making
Knopf et al. ⁴⁹	2008	United States	Autonomy of cohabitants and decision-making

company a higher life expectancy of patients with SCD to improve the quality of life.

The close relationship with chronic illness and the experience with SCD can be understood from some of Schutz's ideas³¹. This author³¹ affirms that each individual builds his/her world. However, this construction does not arise out of nothing. Individuals build from the relationships they establish with other individuals. The world of life is the intersubjective world that precedes each life, and every interpretation about that world is based on a stock of previous experiences²⁰. This intersubjective process can produce a large stock of knowledge that becomes the basis for associative movements of self-help or support for patients³².

The research by Silva et al.²⁸ stands out here, which emphasizes the role of the family as a "care-producing unit". This ideal is highly necessary when analyzing the significant involvement, and the stock of knowledge acquired not only from patients, but also from their families, usually represented by the maternal figure as direct

caregivers, who are clinically aware of the theme, by proposing measures of self-care, and are engaged in the struggle to ensure rights by associative movements.

Barbosa³³ and Motter and Okabayashi³⁴ define patients' associative movements as networks generally formed by patients, relatives, or health professionals who invest in remedying the shortcomings of the conventional health system. The associations of SCD patients play a crucial role in building self-esteem and leadership in the face of illness, and a space for sharing information, experiences, and cooperation³⁵.

Based on Goffman's stigma theory, Moreira and Souza³⁶ observe that associativism integrates people around a cause that involves a stigma. It translates into the relationship between "equals" (people who share the stigma) and "informed" (people who show solidarity with the causes of the stigmatized). Such associations are anchored in the secular knowledge of coexistence and the demands for an expanded social recognition. They can recruit other "informed", "like health

professionals who, because of their technical knowledge, produce a mix in the solidarity/alterity/identification/recognition process³⁶(p.57).

One dimension to be considered in the experience with SCD is race. The link between race and sickle cell disease was motivated by the origin of the disease, which, despite having admitted a multiregional origin, hemoglobin HbS has always been linked to black bodies³⁷, thus configuring a significant dilemma in the theme of sickle cell disease, especially in Brazil and in the United States, making this process of illness and its experiences even more complicated.

Peter Fry³⁸ raises the strong racial significance of the SCD and the persistent role of the black movement in the consolidation of public policies aimed at SCD patients and their relatives. He affirms that the black population's invisibility and lack of responsibility vis-à-vis their bodies were re-signified in the context of SCD, in which campaigns promoted and entrusted blacks with prophylaxis against SCD in the USA, in the 1970s. A more realistic and participatory black community in society was created with this direction of responsibilities. The author also highlights the strong female role in the black movement to ensure SCD-related rights.

Another paper found in this bibliographic search with a racial and gender approach belongs to Reed³⁹, which describes the influence of ethnicity and gender in the process of participation of men in prenatal screening for SCD and thalassemia. The author argues that, despite the lack of all the stereotypes of black parents, members of black minorities were more open to equitable views of gender and great supporters during the gestation of their partners concerning British white men.

Lattimer⁴⁰ addresses the difficulties of hospital care in vaso-occlusive crises in SCD patients, such as delayed drug treatment, overstated expression of pain, and opiophobia and, based on a questionnaire, seeks to quantify the experience of those living with SCD, assuming a significant discrepancy between other patients in hospital care.

While describing a quantitative cohort study, Lattimer's⁴⁰ research was included in this review because it presents an attempt to measure experience. He concluded that SCD patients face more problems during hospital admissions than other patients. The author also suggests that care disparities are due to the ethnic-racial character of those involved, who receive less non-verbal care, empathy, and information during their hospital stay.

SCD as a racialized disease mostly affects black bodies, who are seen and treated as deviants in the face of the white medical authority holder of formal healing knowledge. Addressing ethnic-racial relationships in health for the case of sickle cell disease is of high relevance, mainly when referring to Lattimer⁴⁰, a paper found in this review – the significant care disparity between black patients with SCD in health services vis-à-vis other patients. However, we highlight the need for an intersectional approach to tackle discrimination and chronic illness, which reflects all the criteria of race, gender, and class that are so cohesive in Brazilian society and that show black women living with the disease sickle cell in an extreme and singular condition.

Based on Crenshaw¹⁸, an intersectional analysis of those living with sickle cell disease is strongly recommended, especially when considering the confluence of the categories race, social class, and the chronicity of the disease, as multiple systems of subordination, in the establishment of patients' subjectivities and their support network. The struggle for equity within this circle of oppression is reflected in the achievement of autonomy and participation for those living with the SCD in all decision-making spaces where they still have a peripheral stance.

Participation in clinical research

In this theme, papers addressing SCD patients' participation in clinical research stand out to create new protocols and improve treatment aimed at the disease.

Patterson et al.⁴¹ aimed to identify the benefits and hurdles in the recruitment and participation of patients with SCD in pediatric clinical research. With a similar objective, Lebensburger et al.⁴² identified the obstacles and facilitators in the introduction in clinical research aimed at the treatment with hydroxyurea, with huge debates by the SCD community. Jones and Broome²⁴ also followed an approach to participation in clinical research, which, through the realization of focus groups aimed at African-American adolescents, allowed questioning these patients toward recruitment and participation in research strategies.

Through an ethnography, Benjamin⁴³ brings about the analysis of the relatives' responses regarding recruitment for stem cell transplantation, identifying the uncertainties and concerns regarding this treatment.

Liem et al.⁴⁴ interviewed parents and guardians of children with SCD about consenting to

participate in clinical research and decision-making criteria. Hankins⁴⁵ brings an analysis by Liem et al.⁴⁴, pointing to the complexity of recruiting patients with SCD in a minority context and often in a situation of vulnerability. It also highlights the crucial role of professionals who are sensitive to the socioeconomic context and the family scenario.

Hassell et al.⁴⁶ presented proposals prepared by the American Society of Pediatric Hematology at the SCD Summit for the unification of discourse in clinical and action research. Among the prerogatives pointed out are health care access streamlining, the expanded approaches in basic research, and also the improvement in the role of advocacy, services, and fundraising.

Participation in clinical research can provide SCD patients with access to new technologies, drugs, and highly specific procedures, even in the testing and development phase. Despite the robust ethical criteria and the significant advantages of inclusion in the research of this type, the refusal of cohabitants can be born out of fear of unknown side effects, or even by objectifying the bodies as the focus of the research.

Another substantial obstacle to integrating these patients in clinical research may arise from cultural disparities and difficulties in communication between researchers and the target audience. A flowery language prevails in the academic and medical environment, and technical and specific terms related to diseases and their implications are valued. However, professionals engaged in research are often unable to translate their objectives to potential participants in an enlightening way, generating conflict and refusals⁴⁷.

The studies analyzed here did not reveal the form and context in which participation was requested, which can be considered of extreme relevance for the positive or negative response of cohabitants. When contact is made in a hospital environment, the requesting authority becomes much more imposing, as it is an environment controlled by the medical authority, questioning even the quality of care due to specific responses.

The choice of participating or not in clinical research directly interferes with the autonomy of cohabitants and their leading posture in the face of the illness process and its possibilities for treatments. Taking the lead in the face of care and treatment choices can break the intense cycle of discrimination and stigmas surrounding the disease and build self-esteem to cope with chronic illness.

We expanded the discussion about patient participation in clinical research with some rel-

evant papers found²³⁻²⁷, which present the views of health professionals in the face of sickle cell disease and its processes that can influence both limits about the mentioned participation, and the doctor-patient relationship.

Noke et al.²⁵ explore how health professionals in the United Kingdom decide to test children to identify sickle cell traits. The paper shows that international guidelines do not encourage testing due to possible psychological harm and because it does not present immediate reproductive risks. The authors highlight the reality of the United Kingdom in the face of the necessary debates in favor of SCD and the autonomy of patients and their decision-making. They conclude that there is a lack of standardization, training, and science of international guidelines on the part of health professionals, thus suggesting more significant guidance from these teams, mainly for the recognition of children's rights and their autonomy of cohabitants.

Marsh et al.²³ interviewed 62 residents of Kilifi, Kenya, to consult the population's position concerning the attitude of researchers and health professionals in the face of misaligned parenthood cases involving sickle cell disease. Because it is a genetic disease, SCD's expression occurs through the inheritance of a gene from the mother and another from the father. Thus, the paper discusses the father figures that do not match the genetic SCD makeup. Several questions were raised by the respondents, such as maternal figure exposure, child's disease deterioration, separation/divorce, possible violence against women, among others. The authors conclude that, although the questions have not been clarified, several ethical notes have been elucidated throughout the research, including the need for consultation with the community and all parties involved in the decision-making process.

A second article by Marsh's team²⁴ addresses how researchers should treat information and data on sickle cell disease discovered in research. The disclosure of SCD data was strongly supported by the respondents when the social benefits of this act were duly clarified.

Tsianakas et al.²⁶ address the offering and acceptability of a prenatal care screening program in the United Kingdom to detect sickle cell disease and thalassemia, and the barriers and facilitators of this offering. The authors note the pregnant women's lack of information about SCD and thalassemia, which meant that doctors designated the test as a routine, avoiding women from any possibility of decision regarding screening. Another

obstacle described in the research is the absence of interpreters during visits because they are patients of a non-English first language. As for the facilitators, the doctors' positive attitudes towards the screening program and the pregnant women's desire for healthy children were mentioned.

Bakshi et al.²⁷ present the medical perspective regarding the choice of patients for SCD treatments. The authors mention that doctors specialized in SCD could take two approaches: the collaborative one, which aims at discussing and presenting all the possibilities of treatment, and the proposing, which is anchored to treatment in an attempt to convince the patient to adhere to a particular therapy. These approaches are still permeated by factors such as disease severity, patient characteristics, and institutional approaches. Moreover, the authors emphasize the need and the involvement of cohabitants in the choice process, providing a shared decision regarding the treatment of SCD patients.

The qualification of these health teams is assumed as proposed conformity between health professionals and those living with SCD, seeking a greater understanding of the theme that must transcend the physical and clinical aspects, but that encompasses more human and comprehensive care of patients and their relatives. This integration is fundamental for the relationship between cohabitants and their care and support networks, further contributing to the construction of a positive stance towards the SCD and the involvement of cohabitants in the treatment selection processes.

Through the lens of Goffman¹⁵, the stigma and marginalization of "non-places" imposed on those living with SCD are presented in the papers analyzed very subjectively. Emphasis is placed on the need to involve patients both in clinical research and in the decision-making process as a reflection of the marginalized stance assumed and inculcated to patients in their non-place as a socially stigmatized individual. In this sense, the participation of those living with the SCD can remove or minimize their "non-place", shifting it from invisibility to a leading role.

The autonomy of cohabitants and decision-making

In general, the papers related to this topic address patients' involvement in the decision-making process regarding therapeutic measures and how these patients deal with the treatments presented. When investigating factors that in-

fluence decision-making about SCD treatments, Hankins et al.⁴⁸ observe that the option of cohabitants for drug treatment with hydroxyurea to the detriment of stem cell transplantation or blood transfusion did not cause significant differences between the quality of life of patients concerning the chosen treatment. These authors highlight the importance of children and adolescents expressing viewpoints concerning the treatment and professionals discussing the risks and benefits of each therapy with cohabitants so that shared decision-making is effective.

Knopf et al.⁴⁹ address the preferences of adolescents with SCD in the decision-making process and how compatible the decisions were with that of their parents. These authors consider that patients' involvement in decision-making concerning chronic illness can increase expectations and improve patients' quality of life. They conclude that, despite the substantial benefits of living together, adolescents and parents prefer passive decision-making, influenced by age, parental education, and general health stage.

Besides pointing out the great health care disparities in patients with SCD, Hassel et al.⁴⁶ value community involvement as fundamental for the treatment of this disease. These authors recognize the advantages of including cohabitants in the decision-making around the disease and value health education for these subjects.

Jones and Broome⁵⁰ also highlight patients' autonomy and decision-making as the background for a discussion on the socialization of adolescents in various chronic illnesses. Authors affirm that patients with SCD must have greater control over their treatments compared to diabetic patients analyzed in the same study.

Lattimer⁴⁰ concludes that patients with more information and more involved in the treatment of SCD are satisfied with the doctor-patient relationship, have an improved health status, and a drop in the number of hospitalizations is observed.

The appreciation of the role of SCD cohabitants also reflects in the construction of their subjectivity, considering a comprehensivist approach and emphasizing the individuality and different perceptions and experiences of patients and those who live with sickle cell disease.

The patient's involvement in the treatment decision processes can show an autonomous posture of SCD cohabitants and interfere in the acceptance of the disease, incorporating self-care measures and significant differences into the patient's routine, strongly impacting the illness. For

this, stigmas must be overcome, and the intersubjectivity of those living with SCD and health professionals must be dialogued.

Conclusion

This analysis highlights the dire need for developing research aimed at those living with sickle cell disease. The significant advances in therapies, especially in new drugs, significantly increased the life expectancy of these patients, which increases the uncertainties and insecurities of patients about the little-known future.

It is essential to highlight the vital participation of the SCD subjects in clinical, quantitative, and qualitative research to share experiences and raise the voice of cohabitants as the leading figures of the care process, not only as “target audience” or “research object”. Thus, less hierarchical approaches are suggested, objectively explaining the approaches and benefits of participation, and future contributions and their impact on the participants’ quality of life and the next generations.

Some of the studies analyzed may have stimulated or considered the participation of SCD patients and their relatives with an instrumental

connotation to make research feasible concerning collaboration in recruitment, sample composition, and investigative involvement. Others may have taken advantage of this participation to analyze their findings better or contextualize recommendations to address SCD care. Others enriched their conclusions from the participation of subjects and relatives, shifting them from the simple verification of how the disease was perceived in order to capture the experience of living the disease and, with that – among other inputs – subsidize the health care for people with SCD. In any of the situations, the studies contributed intentionally or not to the greater visibility of the problem that involves being a patient of this disease for the subjects and their relatives.

The themes addressed here, within the context of participation of those living with SCD, are firmly integrated by reflecting the construction of autonomy in the face of therapeutic decisions and the composition of their subjectivities in the face of the SCD.

Finally, there is a need to develop new socio-cultural, qualitative studies that can promote the wide dissemination of their results not only in academic spaces but also to those living with SCD.

Collaborations

WSL Lopes, was responsible for collecting and analyzing the data, writing, and reviewing the manuscript. R Gomes, was responsible for writing, reviewing the paper, and adding significant parts.

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