Revista ARACÊ
ISSN: 2358-2472

SICKLE CELL DISEASE: CLINICAL PROFILE AND DIAGNOSTIC AND DISEASE CONTROL INSTRUMENTS

https://doi.org/10.56238/arev6n4-051

Submitted on: 11/05/2024 **Publication date:** 12/05/2024

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ABSTRACT

The objective of the present study is to present evaluative instruments as a proposal to outline the clinical profile of people with Sickle Cell Disease (SCD). The descriptive study was conducted based on the analysis of clinical trials, reports, guidelines, literature reviews, and available evidence about the most commonly used instruments for measuring pain, quality of life, anxiety, depression, fatigue, and sociodemographic aspects of people with SCD. The clinical profile can be performed through detailed anamnesis with sociodemographic information; pain measurement using the Visual Analogue Scale (VAS) and *Brief Pain Inventory* (BPI); determining quality of life using the *Short Form Health Survey* (SF-36); prevalence of anxiety and depression using the *Hospital Anxiety and Depression Scale* (HADS); and determining the degree of fatigue using the *Patient-Reported-Outcome Measurement Information System – Fatigue* (PROMIS). The search for detailed information on the evaluative instruments used to outline the clinical profile is relevant because it will provide adequate guidance for the follow-up and treatment of SCD.

Keywords: Sickle Cell Disease, Pain, Health Profile.

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ISSN: 2358-2472

INTRODUCTION

SCD is a group of hemoglobinopathies with an incidence of approximately 300,000 to 400,000 births per year worldwide. It is predominantly in black and brown people, with a genetic and hereditary origin, caused by mutation in the HbβS gene. After polymerization, red blood cells take on the shape of a sickle, with a greater predisposition to hemolysis, activation of the immune system, and vaso-occlusion with associated ischemia. These pathophysiological mechanisms contribute to the occurrence of lesions in several organs, especially the brain, kidney, spleen, lungs, heart, and eyes. The diagnosis of SCD is made through neonatal screening tests (heel prick test) and, in adulthood, by electrophoresis examination. As therapeutic approaches that can counteract the severity of SCD, the contribution of prophylactic penicillin and protein-conjugated vaccines to reduce the risk of infections in children with SCD, the use of hydroxycarbamide, which increases fetal hemoglobin and reduces red blood cell sickling, blood transfusions, and hematopoietic stem cell transplantation as a curative therapeutic strategy stand out, although it has limited action as it depends on the availability of donors (Kavanagh, Fasipe, Wun, 2018; Hussein *et al*., 2021).

SCD is accompanied by acute complications – pain crises, acute chest syndrome, and stroke, and chronic complications – pain, kidney disease, nephropathy, cardiac dysfunction, restrictive lung disease, retinopathy, pulmonary hypertension, avascular necrosis of the long bones, iron overload associated with transfusion, priapism, leg ulcers, increased susceptibility to septicemia, among others (Crosby, Quinn, Kalinyak, 2015; Hussein *et al*., 2021). With advancing age, organ damage becomes more frequent, becoming chronic problems, with relevant implications for follow-up and treatment, which makes the pathology a public health problem still poorly understood by health professionals and government entities.

For the care of people with SCD, ideally, early diagnosis, prevention of complications and targeted therapy should be carried out Worldwide, neonatal screening programs have already been implemented, however, challenges related to the evaluation, follow-up, and treatment of complications associated with SCD remain, especially in low-income countries with a high prevalence of this pathology. From this perspective, the biopsychosocial challenges that accompany people with SCD deserve to be highlighted, such as living with chronic pain, the presence of emotional disorders, stigmatization accompanied by institutional racism, reduced quality of life, the tendency to social isolation, fatigue and

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ISSN: 2358-2472

frequent medical consultations that lead to work interruptions, causing personal financial stress and that of the entire family (Hussein *et al*., 2021).

SCD can increase the need for frequent hospitalizations due to pain crises, acute lung injuries, and high risk of infections. For professionals who accompany people with this pathology, it is of fundamental importance to better understand and manage the clinical profile, as well as the presence of associated complications with a view to defining, managing and adopting timely, appropriate and effective therapeutic interventions. Viola *et al*. (2021) point out that advances in health care in recent years have led to an increase of approximately 95% in the transition of children with SCD to adulthood, which reinforces the evolution of general health care for those affected by the pathology. However, the lack of programming for the transition to be effective, as well as the lack of training of professionals involved in monitoring and care, greatly contributes to the high rates of morbidity and mortality among people with SCD.

In this context, studies have been carried out for a better understanding and evaluation of people with SCD, by designing the profile and analyzing the efficacy of clinical trials. However, there are no markers to analyze pain resulting from vaso-occlusive crises specific to this disease. In this sense, Farrell *et al*. (2019) suggest that domains related to acute and chronic pain, emotional impacts, sleep quality, fatigue levels, and social, physical, and cognitive functions should be evaluated.

Although recommendations for approaches have already been proposed, there is no standard for assessing the complications associated with SCD with an emphasis on physical and mental health. Therefore, a greater understanding of the clinical profile is needed through detailed anamnesis with sociodemographic aspects, pain measurement, determination of quality of life, fatigue levels, prevalence of anxiety and depression. Thus, the objective of this research is to present evaluative instruments as a proposal to outline the clinical profile of people with SCD in order to provide an overview of the diagnosis, frequent acute and chronic clinical manifestations and, consequently, to direct best practices for the treatment of SCD.

METHODS

This is a descriptive study based on the analysis of clinical trials, reports, guidelines, literature reviews, and available evidence about the most commonly used instruments for the diagnosis and care of people with SCD. It is noteworthy the performance of a detailed

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ISSN: 2358-2472

anamnesis, duly accompanied by relevant tools aimed at the accurate analysis of pain, through the application of the Visual Analogue Scale (VAS) (Jensen; Karoly; Braver, 1986) and the *Brief Pain Inventory* (BPI) (Keller *et al*., 2004), with special attention to be given to quality of life, which is why the application of the *Short Form Health Survey* (SF-36) (Ciconelli *et al*., 1999) is indicated, associated with the determination of the degree of fatigue through the *Patient-Reported-Outcome Measurement Information System – Fatigue* (PROMIS) (Alves *et al*., 2014), from the study of the prevalence of anxiety and depression based on the *Hospital Anxiety and Depression Scale* (HADS) (Botega *et al*., 1995).

RESULTS AND DISCUSSION

The presence of acute and chronic symptoms and complications of SCD require adequate evaluation to obtain positive responses to the treatment and follow-up of this pathology. According to the treatment-directed Hematology Guidelines for SCD, symptoms should be controlled and complications should be well managed. As treatment strategies, the management of vaso-occlusive crisis and syndromes associated with chronic pain stand out (Maakaron, 2023).

The need for specialized care for chronic pain is highlighted, in addition to the early identification of problems for better guidance to qualified professionals and the support network for affected individuals and their families, with the creation of support for self-management and self-care, as occurs, especially, in priapism, leg ulcers and the use of medications. This conduct reinforces the role of the multidisciplinary team for the adequate monitoring and treatment of people with SCD.

People with sickle cell and their families may face biopsychosocial adversities that include emotional distress, functional deficits, and sociodemographic risk factors, which reinforce the need to identify the individuals most vulnerable to the complications of this pathology. Thus, they can benefit at the appropriate time from the completion of the detailed clinical profile through multifactorial evaluations (Reader *et al*., 2020). Most of the comorbidities associated with SCD need to be monitored in order to avoid the occurrence of physical, emotional, social, and work implications. The detailed anamnesis provides an alternative to get to know the individual more deeply. The evaluation of pain, as it is one of the major complications of SCD, accentuates the possible interferences in biopsychosocial measures related to quality of life, fatigue, anxiety and depression, which show how much

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the disease can influence these parameters. In this context, Chart 1 highlights the instruments most used in the general approach to FD.

Chart 1 – Frequent instruments in the clinical profile of people with SCD

Item	Instruments
Detailed history	Instruments created by researchers with information related to clinical and sociodemographic aspects (Silva *et al*. 2022)
Pain Assessment	Visual Analogue Scale (VAS) (Jensen; Karoly; Braver, 1986) McGill of pain (Pepper; Teixeira, 1996) Brief pain inventory (Keller *et al*., 2004) Pain report (Wilkie *et al*., 2010) *Adult Sickle Cell Quality-of-life Measurement Information System* (ASCQ-ME) (Bulgin; Douglas; Tanabe, 2019) *National Institutes of Health* (NIH) (Litwin, 2002)
Quality of Life Measurement	*Medical Outcomes Study 36 – Item, Short Form Health Survey* (SF-36) (Ciconelli *et al*., 1999) *Adult Sickle Cell Quality of Life Measurement Information System* (ASCQ-Me) (Bulgin; Douglas; Tanabe, 2019) *World Health Organization Quality of Life Brief* (WHOQOL-Bref) (Almarabheh *et al*., 2023)
Prevalence of Anxiety and Depression	*Hospital Anxiety and Depression Scale* – HADS (Botega *et al*., 1995) *PedsQL SCD Module Emotions*(crianças) (Panepinto *et al*., 2013) *ASCQ-ME Emotional Impact* (Bulgin; Douglas; Tanabe, 2019) PROMIS – Depression and Anxiety (Castro *et al*., 2014)
Fatigue Levels	*Patient-Reported-Outcome Measurement Information System – Fatigue* (PROMIS) (Botega *et al*., 1995).

Source: Authors themselves (2024)

DETAILED ANAMNESIS

For the care of people with SCD, it is necessary to carry out a detailed analysis of the history of the pathology, complementary tests in order to collect laboratory data and establish correlations, obtain biopsychosocial information, specify the type and treatments already performed, as well as highlight clinical aspects associated with the pathology such as complaints, signs and symptoms considered general and specific, medications in use, results of medical and cognitive evaluations (Brasil, 2015). The collection of information that is known to be relevant through the anamnestic form ensures the outlining of the sociodemographic profile of the individuals – marital status, level of education according to education levels, profession, number of children, racial self-declaration, monthly income range, type of family residence, among others. Knowledge of the profile of the population, accompanied by the clinical characteristics of the lesions, favors the optimization of treatment, so as to allow specific planning of health care. When discussing evaluation

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options used to trace the biopsychosocial profile of people with SCD, areas for improvement in clinical care will be identified.

In this sense, with the purpose of detecting the aspects that need to be improved in the care of people with SCD in the Republic of Congo, Mukinavi *et al*. (2020) evaluated the knowledge and practice of 460 general practitioners, with an average age of 35 years. It was found that unsatisfactory knowledge about the pathology, associated with failures in the definition of instruments for assessment, diagnosis and treatment, can lead to an increase in morbidity and mortality in people with SCD. It should be noted that the relevant elements related to the conditions for the preservation of citizenship reflect on the confrontation of SCD, aggravated by the precariousness of housing, unemployment, low-paid work and institutional racism. The reversal of these elements can be well assimilated through the adoption of adequate social monitoring policies and effective psychotherapeutic care. The need to promote strategies with the purpose of improving the knowledge of health professionals and the scientific community with a view to better monitoring of this pathology and access to treatment is emphasized.

PAIN MEASUREMENT

The presence of pain in people with SCD is remarkable, affecting 50% of affected adults (Karafin *et al*., 2018), which impacts physical-functional, emotional, social and work losses, negatively reflecting on quality of life. The assessment of pain in SCD is complex and depends on the individuals' reports. The existing assessment instruments contribute in a way to the understanding of the processing and perception of pain. Due to the subjective and multidimensional nature of pain symptomatology, which is peculiar to each individual, assessment instruments are necessary to clarify the triggering mechanisms, intensity, and respective impacts on the individual's life.

In SCD, the signs and symptoms of pain do not always allow an objective measurement, which characterizes it as a subjective manifestation that must be linked to other aspects. By admitting that the evaluation and measurement of pain are essentially objective, there is a risk of not valuing people's voices and not carrying out an empathetic and comprehensive evaluation of those who are in chronic suffering. The objectives of pain measurement should guide a better understanding of the pathophysiology, which fosters the development of innovative therapies and evaluation of the impact of therapeutic strategies with the purpose of reducing suffering (Myrvik *et al*., 2015). As a way of capturing the

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expression of pain in people with SCD through different instruments, the following tools can be used.

It is worth noting that for the multidimensional assessment of pain in SCD in adults, there are health-related quality of life tools that have domains directed to pain and its impacts – *Adult Sickle Cell Quality-of-life Measurement Information System* (ASCQ-ME) (Bulgin; Douglas; Tanabe, 2019), as well as more general instruments such as the *National Institutes of Health* (NIH) (Litwin, 2002). In addition, the McGill pain questionnaire (Pimenta; Teixeira, 1996), the brief pain inventory (Keller *et al*., 2004) and the pain report (Wilkie *et al*., 2010). It is noteworthy that despite being widely used, the numerous pain scales are prone to social influences, psychological factors and possible communication failures that may not be carefully considered (Collins; Renedo; Marston, 2022).

VISUAL ANALOGUE SCALE (EVA)

The instruments most traditionally used to assess pain from a unidimensional perspective in SCD are the Numerical Rating Scale, the Faces Pain Scale, and the Visual Analogue Scale (VAS), in which the individual chooses a single number, face, or line that best represents the intensity of momentary pain in a spectrum at the time the assessment is completed. Farrell *et al*. (2019) indicate the application of VAS appropriately reproduced and administered as an option to measure pain intensity in adults and children over 8 years of age and older, having already been widely used in studies to assess pain in people with SCD (Myrvik *et al*., 2015), revealing efficacy in pain analysis (Dampier *et al*., 2013).

The option to choose VAS is very common in hospitals and outpatient clinics. It is presented in a simple way by means of a 10-centimeter ruler or with the use of a figure numbered in different colors. The VAS consists of a line with the ends numbered 0-10, anchored at the two extremes of pain. At one end of the line is recorded the expression "no pain", while at the other end the definition "worst pain imaginable" is placed. The evaluator asks the individual to analyze and mark the level of pain intensity perceived at the time of the evaluation. The distance between the "no pain" point and the pain intensity indicated by the individual will determine the corresponding value in centimeters on the back of the scale. It is necessary that the participant has visual contact with the scale so that he is able to point out or signal to the researcher the degree of intensity of the pain.

Kazak and Ozkaraman (2021) conducted a randomized controlled interventional study in Turkey, in which they used VAS to measure pain intensity in people with SCD after

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performing exercises with progressive muscle relaxation, in addition to the analysis of coping methods, location, and complaints that accompany pain, a condition that highlights VAS as an important instrument in the analysis of the effects of interventions aimed at improving pain. Tanabe *et al*. (2023), in turn, used VAS in emergency care with the purpose of comparing the time to pain relief among people with SCD with severe pain associated with acute vaso-occlusion episodes, highlighting VAS as a useful tool in urgent and emergency services because it is an accessible and easy-to-apply instrument.

As disadvantages for the application of the VAS, it is evident that: the ability to evaluate the spectrum and the multidimensional nature of the painful experience is limited; there is interindividual variability in the scores found, since the tolerance for pain among individuals can be variable; It requires an additional step in the interpretation of the results found, finally, the need to use paper/pencil or electronic device. Such aspects may make the VAS an inadequate measure for pain analysis when used alone. However, the scores of this scale seem useful in the analysis of outcomes associated with acute interventions according to Kazak and Ozkaraman (2021)

BRIEF INVENTORY OF PAIN

The *Brief Pain Inventory* (BPI) is a short, self-administered questionnaire initially developed for people with cancer, however, it has been increasingly used in other pathologies, with special emphasis on SCD (Dampier *et al*., 2012; Kean *et al*., 2016; Connolly; Hardy, 2019; Abrams *et al*., 2020). The short version of the BPI was developed for use in interventional clinical research with special emphasis on application alongside other multifactorial tests in people with and without painful symptoms (Connolly *et al*., 2019). The instrument describes pain in terms of the intensity of the physical sensation and the influence on function, according to the way pain is described in some generic measures of pain that are widely used, which contrasts with the assessment instruments that include the affective component. The short version contains 15 items, which include 2 scales containing multiple items that include measures for intensity, impact of pain on functioning, and well-being. There are two open-ended questions to measure the location of pain and the type of treatment used for the pain symptomatology that is used by the individual, and a single-item scale to measure the effectiveness of the treatment (Keller *et al*., 2004).

Dampier *et al*. (2012) aimed to find better methods to assess pain associated with SCD in hospitalized adults. To do so, they examined several measures of intensity and

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factors that could measure pain in the upper limbs after comparing drug therapies. They observed that VAS scores measured daily in conjunction with other instruments such as the BPI were sensitive to the change in daily improvements in pain intensity associated with resolution in vaso-occlusive crisis. In the same vein, Kean *et al*. (2016) compared the sensitivity to change and responsiveness after the intervention of pain-related instruments, and found that the BPI measures were more effective in detecting the changes presented by a group of people with persistent musculoskeletal pain of moderate severity, compared to the SF-36 *Bodily Pain instrument*. From this perspective, Osunkwo *et al*. (2021) investigated the relationship between three pain assessment instruments in SCD – visual analogue scale, Kinesiophobia Scale and BPI. They concluded that it is important to carry out objective and subjective pain assessments in order to study it systematically, in a robust and multifactorial way in order to better direct clinical care.

QUALITY OF LIFE MEASUREMENT (SF-36)

The morbidity that usually accompanies SCD is known for its considerable negative impact on the quality of life of those affected (Ballas *et al*., 2006). The *Medical Outcomes Study 36 – Item, Short Form Health Survey*, SF-36 is self-administered and validated by Ciconelli et al. (1999). This instrument consists of 36 questions that seek to assess the health status of individuals in a very comprehensive way, based on evaluation parameters on the level of quality of life of the population under study. The results obtained through the application of pre-established formulas present values attributed to eight domains: functional capacity, physical limitation, pain, general health status, vitality, social aspects, emotional aspects and mental health, which can vary from zero to 100. Results closer to 100 indicate that a better quality of life has been achieved, while results close to zero are suggestive of the opposite.

Numerous studies have evaluated the quality of life after the application of some intervention, especially related to the use of medications in SCD (Ballas *et al*., 2006; Karafin *et al*., 2018). It is known that pain present in SCD usually has the use of opioids as a therapeutic strategy, and in this context, Karafin *et al*. (2018) evaluated the relationship between opioid use and health-related quality of life. Among other instruments, these authors applied the SF-36 to adults with SCD and found that higher doses of opioids are associated with a poorer quality of life, in addition to other predictors such as physical and mental health. Sogutlu *et al*. (2011) examined the impact of the presence of somatic

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symptoms on pain, depression, anxiety, health care utilization and quality of life, through the application of the SF-36 instrument, by 230 adults with SCD. These researchers concluded by associating the presence of pain with depression, anxiety and poorer quality of life.

In the study by Ballas *et al*. (2006), measures of quality of life in health were evaluated in order to verify whether the use of Hydroxyurea presents benefits to 299 individuals. The SF-36 was useful to include pain recall over two years of follow-up and verification that the drug had no apparent effect on measures of quality of life other than pain. In the same vein, Thuret *et al*. (2009) evaluated the individuals' profile, quality of life, adherence and satisfaction with the use of Deferoxamine (iron chelator used in chronic blood transfusions) in 70 people with thalassemia, sickle cell anemia or myelodysplastic syndromes. The SF-36 scores of the individuals evaluated were lower than those of the French population in general, lower among those with comorbidities and those who were dissatisfied with the treatment, which suggests that quality of life was compromised by adherence or not to the proposed treatment.

Another instrument widely used to assess the quality of life of people with SCD is the *Adult Sickle Cell Quality of Life Measurement Information System* (ASCQ-Me) (Bulgin; Douglas; Tanabe, 2019). It is a tool developed in 2002, designed specifically for SCD, which aims to provide a systematic, valid and reliable means to document self-reported results of adults with SCD. The ASCQ-Me evaluates quality of life according to the following domains: emotional impact, pain episodes, pain impact, medical history, impact on sleep, impact on social functioning, and impact of joint stiffness, according to Keller *et al*., 2014.

Also noteworthy is the *World Health Organization Quality of Life Brief* (WHOQOL-Bref), an evaluation instrument of quality of life of the World Health Organization, with high internal consistency and validity in the assessment of the quality of life of individuals with SCD (Almarabheh *et al*., 2023). By using this instrument, Roberti *et al*. (2010) evaluated the quality of life of 60 individuals with SCD, aged between 14 and 60 years, undergoing treatment at the Hospital das Clínicas of the University of Goiás. They applied the WHOQOL-Bref and found scores (from 0 to 100) of 57.32 in the physical domain, 66.03 in the psychological domain, 69.86 in the social domain and 52.76 in the environmental domain. These findings demonstrated a significant correlation between prejudice due to the disease and educational level, suggesting that SCD can limit the survival of the individual with the impairment of quality of life.

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It is notorious that several researchers around the world have used the SF-36 with the intention of evaluating the health-related quality of life of people with SCD (Ballas *et al*., 2006; Sogutlu *et al*., 2011; Kean *et al*., 2016; Karafin *et al*., 2018), since the complications that are associated can have a negative impact with repercussions on physical and mental health and are often an aspect neglected by health professionals. It is important to highlight that the assessment of quality of life in SCD can help in the general prognosis and in a better targeting of interventions with the purpose of providing excellence in health care and, consequently, increasing the life expectancy of these individuals, in view of the way complications and financial burdens are imposed by this disease.

ASSESSMENT OF ANXIETY AND DEPRESSION (HADS)

The Hospital *Anxiety and Depression Scale* (HADS) is a brief investigative questionnaire that is considered simple for the individual to understand. It is a tool capable of providing information about anxiety and depression levels. Its Portuguese version was validated by Botega *et al*. (1995) and aimed at the evaluation of people with chronic pain by Castro *et al*. (2006). Several studies use the HADS scale to screen anxiety and depression in individuals with SCD (Olowoselu *et al*., 2021; Oliveira *et al*., 2023), although there is a recommendation for the use of the PedsQL SCD *Module Emotions* for children and the ASCQ-ME *Emotional Impact measure* for adults, PROMIS measures – Depression and Anxiety (Farrel *et al*., 2019),

The HADS scale consists of 14 questions, seven of which are related to the level of anxiety (odd) and seven related to signs of depression (even). Each option, of the four possible answers related to all questions, has an assigned value (0, 1, 2 or 3) that allows the totalization of a score for anxiety and another for depression as follows: 0 - 7 points = unlikely, 8 - 11 points = questionable or doubtful (possible) and 12 - 21 points = probable. Regarding the limitations of other psychometric tools, the use of HADS is recommended to avoid overlap between symptoms of depression and those of sickle cell anemia (Oudin *et al*., 2021).

Castro *et al*. (2006) in the validation study of the HADS scale, they carried out a cross-sectional descriptive study through the evaluation of 91 individuals from the Pain Center through the application of interviews using the HADS Scale and *the International Neuropsychiatric Interview Brazilian*. Through the HADS, the results showed that approximately 60% of the respondents had anxiety and about 40% had depression. In a

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descriptive cross-sectional study, which was part of a randomized crossover clinical trial, Oliveira *et al*. (2023) evaluated 131 adults with SCD in the state of Bahia, Brazil, using HADS. Of the 131 individuals approached for participation, 75 completed all questionnaires with HbSS and HbSC genotypes. Of these, 49 (65.3%) were women, with a mean age of 34.13±10.02, while 66 (88.0%) self-declared as black. The mean severe pain was 3.86±2.74; 40 (53.33%) had anxiety, 25 (33.33%) had depression, and 15 (20.0%) reported having suicidal thoughts.

Olowoselu *et al*. (2021) determined in Nigeria in 226 people with SCD, the prevalence of depression and anxiety and their respective associations with pain, having verified the predominance of 49.5% and 30.5%, related to symptoms of anxiety and depression, respectively. In addition, approximately 70% and 46% of people with pain had abnormal scores for anxiety and depression. This phenomenon reflects a high prevalence of these symptoms associated with pain as an important predictor for the occurrence of disorders of this nature.

EVALUATION OF FATIGUE (PROMIS)

Fatigue is a common symptom in people with SCD. Although the exact pathophysiology that results in fatigue is not fully understood, it is hypothesized that it may be secondary to the underlying abnormal processes of sickle cell cells, including ongoing hemolysis, vaso-occlusive events, pain, and anemia. PROMIS is a test that aims to inform self-reported symptoms, from a slight subjective feeling of tiredness to a feeling of overwhelming, debilitating and sustained exhaustion. PROMIS forms are universal, not pathology-specific, and are designed to be applied worldwide. It is one of the four measurement instruments belonging to the *Person-Centered Assessment Resource* (PCAR) (Keller *et al*., 2017), with few studies in people with SCD. Most of the instruments assess the fatigue experienced in the last seven days, taking into account that it is a common symptom in people with SCD that certainly reduces the person's ability to perform daily activities and perform family or social functions normally. Although the pathophysiology indicates that fatigue is not fully understood, it is hypothesized that it may be secondary to the underlying abnormal processes of sickle cell red blood cells, which include continuous hemolysis, vaso-occlusive events, pain, and anemia.

In this sense, Katharine *et al*. (2023), in a systematic review study, sought evidence to understand the use of PROMIS – fatigue in studies of SCD populations. The PROMIS

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scores reported in these studies seek to characterize fatigue estimates across the entire evidence base in order to assess the usefulness of this tool as a measure of fatigue in this population. They observed that despite the weight of fatigue in people living with SCD, the results of this research demonstrated that tools such as PROMIS – Fatigue are little used and, when used, are implemented inconsistently. The PROMIS – Fatigue scores available in people with SCD were marginally worse than in the general population, however, the high degree of heterogeneity makes it difficult to draw solid conclusions from the available data (Batt *et al*., 2023). Other studies have analyzed individuals with SCD and used PROMIS – Fatigue as an assessment tool, such as LaBedz *et al*. (2022), Curtis and Brandow (2017), Keller *et al*. (2014).

In view of the above, it is worth noting that the dissemination of awareness about SCD to the entire community is important for the successful implementation of planned programs. Government funding and support for community education should be sought, as well as facilities for implementing the programs (Babalola *et al*., 2019). It is known that the ideal is to maintain maximum autonomy and independence to ensure a good quality of life (Mendonça *et al*., 2024). However, for people with SCD it is difficult to learn to live with this pathology, particularly young adults. A biopsychosocial and multidisciplinary approach is needed for good care and follow-up of these individuals. All team professionals must collaborate in a coordinated manner as they seek to understand the interaction between biological, psychosocial and sociological factors in SCD with the purpose of improving the general health status of those affected by this pathology (Crosby, Quinn, Kalinyak, 2015).

The limitations of the study include the fact that it was essentially based on data from other studies. Longitudinal studies and clinical trials with more specific instruments for SCD in different clinical settings are needed. However, the present review contributes to support the scientific literature, since it has SCD as a reference. The improvement of multifactorial knowledge reaffirms the relevance of the exchange of experiences among peers, always from the perspective of interdisciplinarity. Thus, the objective of this proposal is fulfilled, which is to present evaluative instruments as alternatives to outline the clinical profile of people with SCD, which can contribute to recover the health of citizens, always with the purpose of achieving an improvement in the general state of health and, consequently, in the quality of life. Certainly, the results of this study may offer new subsidies to the agencies responsible for the implementation of public health policies, with emphasis on those that focus on SCD. In this way, it will contribute to advance in the elaboration of strategic plans

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that have in training and updating programs, multidisciplinary health alternatives that increasingly strengthen the relationship between health professionals and society.

CONCLUSION

The application of specific instruments can measure the various elements related to sociodemographic aspects, pain, quality of life, prevalence of anxiety and depression, and fatigue levels of people with SCD. The rigorous use of specific instruments can make the profile of people with SCD reliable, therefore, capable of leveraging initiatives that provide solutions that can be tested and properly associated with the effects of therapeutic proposals aimed at the study of SCD. After evaluating the clinical profile of people with SCD, it is possible to develop investigation, follow-up and treatment protocols, particularly pharmacological, which can homogenize the care of people with SCD to be treated in health services. It is worth noting the possibility that the proposed instruments can count on the participation of public institutions, entities focused exclusively on research and development, public and private agencies, always from the perspective that the alternative of these instruments is based on scientific bases. In this way, the instruments may be available to institutions, since they have the potential for the elaboration of educational campaigns aimed at health promotion and prevention and planning aimed at SCD in response to this demand from society.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest in the publication of this article.

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ISSN: 2358-2472

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