

# Being an adolescent despite the restrictions and discrimination imposed by sickle cell disease

Ser adolescente apesar das restrições e da discriminação impostas pela doença falciforme

Ser adolescente pese a las restricciones y a la discriminación

impuestas por la enfermedad de células falciformes

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## Abstract

**Objective:** Understand the experiences of being an adolescent with sickle cell disease.

**Methods:** Qualitative study, conducted at a reference service in the state of Bahia, Brazil between March and June 2018. Ten adolescents with sickle cell disease participated. The data were obtained through drawings-and-stories with a theme and semi-structured interviews and analyzed based on Grounded Theory.

**Results:** The experience of adolescents with sickle cell disease is represented by the core category of “trying to be a normal adolescent, despite the restrictions and discrimination the sickle cell disease imposes”, and five other categories: “Feeling different from other adolescents”, when they see that their growth has altered, that they experience medical problems and compare themselves to other adolescents, “Living with restrictions in their daily routine”, of physical and food-related restrictions, required in self-care and disease management to achieve quality of life, “Living in bad situations”, in the experience of pain, constant hospital visits, fear of death and uncertainty about the future, “Feeling like a normal adolescent”, when they were able to maintain their social activities, including school, friends, and family, and “Realizing the stigma”, when they fear the discrimination and adopt ways to hide that they have the disease.

**Conclusion:** By seeking to be a normal adolescent, the participants aspired to take control over their own lives, avoid breaks from the routine and meet social expectations, protecting their identity from labels and discrimination.

## Resumo

**Objetivo:** Compreender as experiências de ser adolescente com a doença falciforme.

**Métodos:** Estudo qualitativo, realizado em unidade de referência no estado da Bahia entre março e junho de 2018. Participaram dez adolescentes com doença falciforme, os dados foram obtidos mediante desenhos-estória com tema e entrevistas semiestruturadas e submetidos à análise embasada na Teoria Fundamentada nos Dados.

**Resultados:** A experiência do adolescente com doença falciforme é representada pela categoria central “Buscando ser um adolescente normal, apesar das restrições e da discriminação impostas pela doença falciforme” e mais cinco categorias: “Sentindo-se diferente dos outros adolescentes”, ao perceberem seu crescimento alterado, vivenciarem problemas clínicos e se compararem aos demais adolescentes; “Vivendo com restrições em sua rotina diária”, de ordem física e alimentar demandadas no autocuidado e manejo da doença para o alcance de qualidade de vida; “Vivenciando situações ruins”, na experiência de dor, constantes hospitalizações, medo da morte e incerteza quanto ao futuro; “Sentindo-se um adolescente normal”, quando

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podiam manter suas atividades sociais com escola, amigos e família; e “Percebendo o estigma”, ao temerem a discriminação e adotarem modos de ocultar que possuíam a doença.

**Conclusão:** Ao buscar ser um adolescente normal os participantes aspiraram assumir o controle sobre a própria vida, evitar rupturas na rotina e atender às expectativas sociais, protegendo sua identidade de rótulos e discriminação.

## Resumen

**Objetivo:** Entender las experiencias de ser adolescente con la enfermedad de células falciformes.

**Métodos:** Estudio cualitativo, realizado en una unidad de referencia en el estado de Bahia entre marzo y junio de 2018. Participaron diez adolescentes con la enfermedad de células falciformes, se obtuvieron los datos por medio de dibujos-historia con tema y entrevistas semiestructuradas y sometidos a análisis con base a la Teoría Fundamentada en Datos.

**Resultados:** La experiencia del adolescente con enfermedad de células falciformes está representada por la categoría central “Busca ser un adolescente normal, pese a las restricciones y a la discriminación impuestas por la enfermedad de células falciformes” y otras cinco categorías: “Se sienten diferentes de los demás adolescentes”, cuando se dan cuenta de su crecimiento alterado, vivencian problemas clínicos y se comparan a los demás adolescentes; “Viven con restricciones en su rutina diaria”, de orden físico y alimentario que se demandan en el autocuidado y en la gestión de la enfermedad para lograr la calidad de vida; “Vivenciando malas situaciones”, en la experiencia del dolor, constantes ingresos a hospitales, miedo a la muerte e incertidumbre con relación al futuro; “Sentirse un adolescente normal”, cuando podían mantener sus actividades sociales en la escuela, amigos y familia; y “Percatándose del estigma”, al sentir temor de la discriminación y adoptar formas de ocultar que padecen la enfermedad.

**Conclusión:** Al intentar ser un adolescente normal, los participantes aspiraron a asumir el control sobre sus propias vidas, evitar rupturas en la rutina y atender a las expectativas sociales, protegiendo su identidad de clasificaciones y de discriminación.

## Introduction

It is estimated that, in Brazil, each year, about 3,500 children are born with sickle cell disease (SCD) and 200,000 carriers of the sickle cell trait. Although it is distributed throughout the territory, the highest prevalence rates are recorded in the states of Bahia, Rio de Janeiro, Pernambuco, Minas Gerais, and Maranhão.<sup>(1)</sup>

This is a group of hemoglobinopathies caused by a mutation in the genes of the beta-globin chain and identified by the presence of hemoglobin S. This mutation induces the polymerization of hemoglobin, causing deformation of red blood cells and, consequently, ischemia by repeated vaso-occlusive episodes, chronic inflammation, and hemolysis that lead to multiple organ damage and a multitude of complications.<sup>(2)</sup>

The first signs and symptoms of SCD include swelling of the hands and feet; symptoms of anemia, with fatigue or extreme tiredness; and jaundice. Over time, SCD can lead to complications such as infections, growth retardation, and acute pain episodes.<sup>(3)</sup>

In adolescence, a phase of life marked by intense physical, emotional, hormonal, social, and psychic transformations, chronic diseases such as SCD will potentiate bodily changes that will affect the emotional state, self-image, and social development of adolescents.<sup>(4,5)</sup>

The wide variety of complications requires constant primary care and follow-up from many specialties, including hematology, pulmonology, nephrology, orthopedics, pain control, and psychiatry.<sup>(6)</sup> To reduce symptoms and complications, increase survival and improve quality of life, parents need to assimilate a strict care regime and the sick person needs to maintain it throughout life, through self-care measures. Adolescence is considered a time of transition from being cared for to being autonomous for self-care.<sup>(7)</sup>

Thus, after the diagnosis of chronic disease, the changes the disease and its treatment cause start to guide the life of the adolescents and their family members. This journey is long, painful, and often permeated by difficulties and feelings represented by anguish and uncertainties.<sup>(8)</sup> The care demands of a teenager with chronic disease are diverse, highlighting hygiene, physical appearance, medication use, nutrition, and modified habits, requiring the constant ability to rearrange, renew and rebuild their way of life.<sup>(9,10)</sup>

In a search conducted in the SciELO, PubMed, and Cochrane databases in March 2018, using the descriptors “adolescent and sickle cell disease”, studies were found focusing on the prevalence of complications of SCD and its clinical management, psychological therapies for pain control, and family caregivers’ experiences. These findings indicated the

lack of qualitative studies that considered the disease experience in the adolescents' narratives.

Knowing that the chronic illness processes involve meanings, perceptions, and self-management of the adversities that the disease promotes, to direct therapeutic plans that reach the ways of being of adolescents with SCD, the question that guided this article was: "How do adolescents experience this phase of development, having SCD?". This study aims to understand the experiences of being a teenager with SCD.

## Methods

Qualitative, exploratory, and descriptive study, which used symbolic interactionism as a theoretical framework, as it is a perspective of analysis of human experiences, focusing attention on the interaction between people who, after interpreting and defining the situations experienced, act in the social context they are inserted in.<sup>(11)</sup> As a methodological framework, Grounded Theory was used.<sup>(12)</sup>

The study was developed in the community room of the Municipal Reference Center for People with Sickle Cell Disease (CMRPDF) located in Feira de Santana, Bahia, Brazil. For this research report, the recommendations of the guide *Consolidated Criteria for Reporting Qualitative Research* (COREQ) were followed.

Ten adolescents participated, included in the study because they had a confirmed diagnosis of SCD. The exclusion criteria were: presenting symptoms, discomfort, or complications at the time of the meeting. The adolescent age group was defined according to the Statute of the Child.<sup>(13)</sup> The data were produced in the community room of the reference center, where researchers and participants were already taking part in joint activities in the matrix project "Representations About the Body and Sickle Cell Disease: Repercussions on Everyday Life, Care, and Sexuality".

The data were collected through the application of the projective technique of drawing-and-story<sup>(14)</sup>, followed by semi-structured interviews conducted by two researchers. The interview script contained

data on the adolescents' demographic characteristics (age, sex, ethnic origin/color) and on the SCD (type of hemoglobinopathy and time since diagnosis), as well as open questions about what it is like to live with SCD and the experiences of a teenager with this condition. The conversations were held in a closed and private environment at the health service and at home, where the participants felt comfortable to express their opinions, with the support of the psychologist, who was in an adjacent room. The mean length of the interviews was 15 minutes.

The adolescents were aged between 12 and 15 years. Six were male and four were female. Five declared themselves black and five mulatto. As for the type of hemoglobinopathy, six had HbSC and four had HbSS. For the majority, SCD was discovered during neonatal screening. The most frequently cited complications were pain crises, infections, and anemia.

The testimonies and contents of stories were recorded and subsequently transcribed in full. They were analyzed according to the Grounded Theory (GT) analysis.<sup>(12)</sup> Following the GT steps, the production and analysis of the data occurred simultaneously. During the open coding, the material was analyzed line by line, apprehended, compared for similarities and differences, contrasted and categorized, to identify the substantive codes.

In the axial coding, the substantive codes were regrouped to form explanations about the phenomena under investigation and to permit the emergence of categories.<sup>(12)</sup> To establish relationships between the elaborated categories and identify the substantive category, the paradigmatic model was used, based on the following components: phenomenon, context, causal and intervening conditions, strategies, and consequences.<sup>(12)</sup>

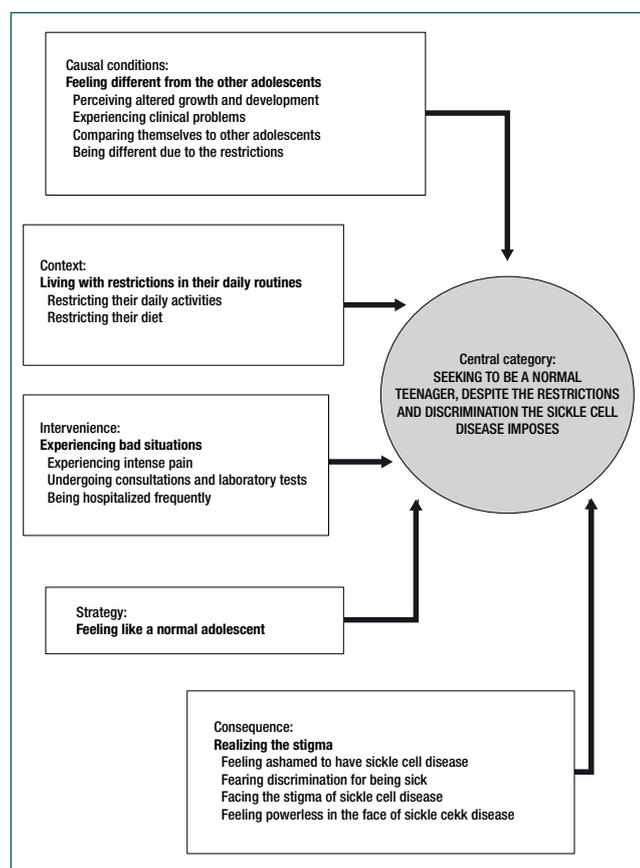
In the selective coding, the categories were integrated and refined into an analytical model, which consists of defining the central category and then describe the concepts regarding properties and dimensions.<sup>(12)</sup> The central category was called "Seeking to be a normal teenager, despite the restrictions and discrimination the sickle cell disease imposes".

The participants received the interview transcripts, drawings, and empirical categories for vali-

dition. In all stages, the ethical standards for research involving human beings were respected. Approval for the research was registered under Opinion 1.440.239 and CAAE 49493315.3.1001.0053. The adolescents signed the Free and Informed Assent Form and their responsible caregivers signed the Free and Informed Consent Form.

## Results

The experience of being an adolescent with SCD can be understood based on the central category, represented in Figure 1, and the synthesis of the adolescents' statements in Charts 1 and 2.



**Figure 1.** Central category “Seeking to be a normal teenager, despite the restrictions and discrimination the sickle cell disease imposes”

In the category “Feeling different from other adolescents”, the perception of the difference from those who did not have their condition was expressed in the recall of situations, such as episodes

of vomiting, lung problems, and injuries that were difficult to heal, which required long periods for recovery, in addition to frequent visits to reference health services.

Adolescents with SCD were unable to engage in leisure or other basic activities, as these demand greater efforts and can trigger pain crises, fatigue, and tiredness. Then, they realized that their lives were limited and restricted. The adolescents felt separated from their social context, in which they had to abandon pleasurable practices, making them self-affirm as “different”, to the extent that they took distance from the activities of their peer group, mainly during sports and physical education. This perception situated them as unequal because they saw themselves as separated and unable to reach this “other” teenager, due to the restrictions family members often imposed. As a result, they wanted to surpass their limitations, despite knowing of the consequences it would entail for their health.

The category “Living with restrictions in their daily routine” showed the physical and dietary restrictions demanded in self-care and disease management to achieve quality of life. In this category, the adolescents highlighted the tiredness when practicing leisure and physical exercises, due to the signs and symptoms of the SCD itself, leaving them idle and indoors most of the time.

Due to the dietary adjustments, the adolescents sadly realized these limitations, because they could not eat what they liked and had to follow a moderate diet. Their compliance with the dietary restrictions was motivated by the fear of precipitating a painful event or crisis due to certain foods.

The adolescents mentioned the parents’ or relatives’ requirements as a form of care and protection, so that they would not suffer from complications, and also as an obligation, as they were not allowed to eat everything they wanted or their favorite foods. The adolescents often declared that this care was excessive and affected their freedom concerning their diet.

The category “Experiencing bad situations” was anchored in the experiences of the barriers for the patients’ lives. They expressed feelings and questions when comparing themselves with their peers

without the disease, anxieties, and fears related to the painful experience, its duration, and recurrence. The pain experience made them feel distressed and uncomfortable, as well as different from other hospitalized patients, who seemed not to suffer as much pain as they did.

Undergoing routine consultations and procedures was seen as a tiring, repetitive and prolonged process. The daily consultations and, often, the surgical procedures led these adolescents to experience physical and emotional stress, generating sadness and discomfort for being often subjected to disease evaluations and controls.

The adolescents with SCD were afraid of possible complications and hospitalizations, as these

implied interrupting their interests to spend an unpredictable length of time in the hospital, disorganizing their daily routine, besides causing anxiety due to the possible discovery of new complications.

The category “Feeling like a normal adolescent” presented the strategies the adolescents with SCD adopted to face the substantive category. It revealed the resources they use to assert themselves like the other adolescents without this condition but, at the same time, it highlighted the extent to which they recognized themselves with additional care needs to sustain life. The restriction of freedom and limitations reinforced the desire to have a life closer to what they considered normal. Thus, they sought to practice activities equal to those of their peer group,

**Chart 1.** Synthesis of adolescents' statements (part 1)

Synthetic picture of adolescents' statements
<p><b>Causal conditions:</b>  <b>FEELING DIFFERENT FROM THE OTHER ADOLESCENTS</b>  <b>Perceiving altered growth and development</b>  <i>A have an 11-year-old colleague who is taller, stronger, more developed than me. Others who do not have the disease evolve faster than those who have the disease. (Scoob, 11 years old).</i></p> <p><b>Experiencing clinical problems</b>  <i>If the person gets hurt, it creates that very big injury, then the whole leg has to be isolated. I had an injury here in my leg that needed repeated dressings at the CSU [health service]. I kept the leg injury for three months. (Manchester Black, 13 years old).</i>  <i>I've tried to play. I played as a winger, but then I stopped, because I saw that it was not working anymore, because the legs hurt too much [...] Everything is bad, I cannot go out in the damp night air, I cannot go out at night [...] (Messi, 11 years old).</i></p> <p><b>Comparing themselves to other adolescents</b>  <i>Adolescents without the disease can play soccer, they can do everything, they can do whatever they want. Those who have the disease are isolated from playing ball. If you play ball, your legs hurt; if you run, your legs hurt; if you play capoeira, your legs hurt; if you play basketball, your arms hurt. Everything, any sport, hurts. (Manchester Black, 13 years old).</i>  <i>A normal teenager [...] she doesn't feel much in the lungs, while adolescents with sickle cell disease feel a lot in their lungs [...] (Wonder Woman, 10 years old).</i></p> <p><b>Being different due to the restrictions</b>  <i>There are things they [adolescents without sickle cell disease] can do and then I don't. Like walking a lot. My friends like to go out, walk and sometimes I do not go out with them because of sickle cell disease, because I cannot get too tired [...] It's bad because I'd like to do pretty much everything and there are things I can't do. (Di Monaco, 13 years old).</i>  <i>Only once my grandmother did not want me to do physical activity, because it would harm me, and I felt different because I could not participate [...] Sometimes my mother tells me not to get too much weight, not to exercise too much, that it can hurt. (Di Monaco, 13 years old).</i></p>
<p><b>Context:</b>  <b>LIVING WITH RESTRICTIONS IN THEIR DAILY ROUTINES</b>  <b>Restricting their daily activities</b>  <i>In games, I get tired faster than other people [...] when I'm playing with my friends, we're playing, I'm already tired and they're not. (Scoob, 11 years old).</i>  <i>You can't play ball. You always have to go to the doctor [...] (Messi, 11 years old).</i></p> <p><b>Restricting their diet</b>  <i>[...] I feel like, like fear of eating something and feeling bad, or having some bad pain there [...] I cannot eat the pig, because I can feel some pain, because of the meat [from the pig] because like my father feels [...] he tells me not to eat it either. Then I keep thinking that, if I eat it, I might feel some pain. (Goku, 14 years old).</i></p>
<p><b>Interveneance:</b>  <b>EXPERIENCING BAD SITUATIONS</b>  <b>Experiencing intense pain</b>  <i>It hurts, man! It hurts everywhere [...] it hurts in the back, arms, legs, even the knee... it hurts everywhere, you have no idea [...] if I could control it, that's fine, but it hurts, it hurts, it feels like your arm is going to crack, that something is going to happen on the outside, your bones look like they are swelling, your arms look like they are broken, it hurts a lot! If you had it, you'd know how it hurts. (Manchester Black, 13 years old).</i>  <i>I feel the pain that hurts in the legs, in the arm, in the spine. The gallows, that is, the pain that we feel, that he is feeling when he is hanged, and that is the pain that I feel because I have sickle cell anemia [...] I lack the courage to do anything, I feel incapable. (Manchester Black, 13 years old).</i></p> <p><b>Undergoing consultations and laboratory tests</b>  <i>He doesn't get examined like I do either [...] I feel bad because it bothers me to have to go out to go to the doctor, get tested. I don't like it very much [...] it's a bad thing because no disease is good, is it? (Goku, 14 years old).</i></p> <p><b>Being hospitalized frequently</b>  <i>It's something more or less. The disease keeps bringing the person straight to the hospital. In that case, it's bad. But also, sometimes, she takes her to the hospital and finds out that he's got something else without knowing anything. (Scoob, 11 years old).</i></p>

**Chart 2.** Synthesis of adolescents' statements (part 2)

Synthetic picture of adolescents' statements
<p><b>Strategy:</b>  <b>FEELING LIKE A NORMAL ADOLESCENT</b>  <i>I think that because I have the disease, it does not mean that I am not normal like others, just that I am a little different, with more care [...] a little more care than other people [...] even if I have sickle cell disease, that doesn't mean I can't do the other things like a person who doesn't have it. (Ana Karla, 15 years old).</i>  <i>I feel normal. Nobody is perfect! Each with his flaws, but I have nothing to talk about no. I've never felt great pain because of it. It has never got in my way [...] I have never stopped doing anything because of it. I do everything I want to do. It's all normal for me. (Goku, 14 years old).</i></p>
<p><b>Consequence:</b>  <b>REALIZING THE STIGMA</b>  <b>Feeling ashamed to have sickle cell disease</b>  <i>At school, my friends don't know, because I don't count, because I'm afraid they'll let me go, that they won't want to play with me anymore [...] I think that, because I have sickle cell disease, my friends will isolate me. (Manchester Black, 13 years old).</i>  <i>My friends do not know that I have the disease. I didn't tell [...] I'm ashamed [...] I had a girlfriend and never told her either, because I was ashamed, because it's a bad disease [...] that has no cure [...] so I didn't want to tell. (Messi, 11 years old).</i></p> <p><b>Fearing discrimination for being sick</b>  <i>I'm ashamed to have sickle cell disease [...] you have a disease, your friends find out and you will no longer be able to walk with them; you have to remain isolated. (Manchester Black, 13 years old).</i>  <i>I feel that because they don't have the disease, they may feel better than me. Just because I have the disease, I cannot do that, and also, sometimes, the person thinks that I, with the disease, will not manage. (Scoob, 11 years old).</i></p> <p><b>Facing the stigma of sickle cell disease</b>  <i>I am not ashamed, because I think it is a good thing and there is nothing bad in the disease [...] It is not a disease to hide [...] I'm not ashamed to tell anyone no, but my girlfriends never knew and none of my friends know that I have it, but I'm not ashamed of anything, because it's not a communicable disease or anything like that. We just don't talk about it. (Goku, 14 years old).</i>  <i>I've had a boyfriend and he knew about my illness. I told it myself and my friends also know, but they treat me normally. (Ana Karla, 15 years old).</i></p> <p><b>Feeling powerless in the face of sickle cell disease</b>  <i>I don't think I'm going to be able to study and work; I'm not going to be able to do anything because of sickle cell disease. Sometimes I feel very bad about it; I feel sad because it limits me. (Ana Karla, 15 years old).</i></p>

to affirm this normality and demonstrate that they were equal to an adolescent without the disease.

Despite the adversities, adolescents with DF, in a positive perspective, recalled what they were capable of accomplishing like the other adolescents without the disease. When the disease did not affect or interfere with their life or their routine and activities, they stated that they felt the same as other adolescents and stressed that, without this self-affirmation, they would be excluded, marginalized, and discriminated against because they had the disease.

Positively affirming the differences was a protection strategy against stigma, because, for the interviewed adolescents, the disease experience made them different and limited. They considered this to be part of human existence and recognized themselves as singular and normal adolescents like any other though.

The category “Perceiving the stigma” showed that adolescents with SCD felt shame. They felt different from other people, because they lived with a chronic disease, which conditioned them to see themselves as disabled people, and feared being judged in a derogatory way.

The fact that the disease is little known, is incurable and causes exclusion motivated them to hide their condition because they fear discrimination.

For them, diseases, in general, were not subjects of interest, commented on, or discussed among adolescents. Therefore, they used the strategy of “hiding” their health condition, seeking a form of protection of the integrity of their identity, preserving themselves, and adopting silencing. When they noticed that family and friends supported them and that their social circle accepted their health condition, however, the coping strategy ceased to be concealment and became disclosure, leading them to talk more about their condition and disease, aiming for others to understand them.

The repeated bouts of widespread pain that disabled them made them fear death and reflect on life. Concern for the future and the feeling of sadness arose because they believed that the SCD compromised their plans and aspirations. Thus, the adolescents' experience, marked by doubts and uncertainties, led them to perceive themselves as powerless in the face of their health condition.

## Discussion

The limitations of this study were the low participation of girls, which made it impossible to infer the experiences marked by gender, combined with

the fact that it was applied at a health service, with the parents' prior consent, an aspect that may have inhibited the participants from addressing issues related to the sexual-affective dimension, because they feared that the responsible caregivers would have access to the data.

This study contributes to broadening the knowledge about group behavior towards chronic illness and provides subsidies for the implementation of measures to reduce the vulnerability of adolescents with SCD. Thus, investments in the disclosure about the disease, in the contexts the adolescents attend, may foster positive strategies to live with the disease.

The category "Feeling different from other adolescents", considered the cause of the central category, was also observed in another Brazilian survey of adolescents diagnosed with chronic kidney disease.<sup>(15)</sup> This category was supported by "Perceiving altered growth and development", "Experiencing clinical problems", "Comparing oneself to other adolescents" and "Being different due to the restrictions".

SCD affects adolescents' growth and development. When they identify that their peers and friends are taller and more robust, they perceive that these milestones in their lives differ from other adolescents. In addition, chronic illness causes physical changes, either explicitly or implicitly,<sup>(15)</sup> such as leg ulcers and jaundice, which directly affect the body image, causing worry, sadness, isolation, and depression.<sup>(15,16)</sup> This confrontation with the chronic disease begins when the adolescents realize that there are differences between their daily routine and that of their friends and family, such as not being able to play.<sup>(4)</sup>

The context in which the central category "Seeking to be a normal teenager, despite the restrictions and discrimination the SCD imposes" was developed was represented by the category "Living with restrictions in their daily routines", constituted by the subcategories "Restricting their daily activities", in which adolescents found themselves living with restrictions in the practice of physical activities, due to the discomforts associated with SCD, and "Restricting their diet". The latter was also ob-

served in a study involving adolescents diagnosed with kidney disease who required some form of dialysis therapy.<sup>(15)</sup>

Thus, the experience of adolescents with SCD was permeated by deprivations, which were manifested by the inappropriate performance and/or non-performance of occupational roles expected for this phase of life. These deprivations could lead to unsatisfactory quality of life and have negative repercussions in the future, affecting their treatment compliance and generating deficits in health and well-being. These factors could lead to complications and major aggravations arising from the disease.<sup>(17)</sup>

The category "Experiencing bad situations", consisting of the subcategories "Experiencing severe pain", "undergoing consultations and laboratory tests" and "Being hospitalized frequently", was outlined as the intervenient condition, in which painful crises stood out in terms of the difference between adolescents with SCD and others without this chronic health condition.

In addition, the interviewees revealed the changes in their lifestyle, evidenced by the changes in daily activities resulting from treatment, in line with the literature.<sup>(15)</sup> They cited, as an example, the distance from school activities, from living with friends and colleagues.<sup>(18)</sup> Studies report that adolescents with SCD need to regularly attend health services,<sup>(15,19)</sup> they are hospitalized, use medication continuously, besides undergoing diagnostic and therapeutic procedures as the disease progresses.<sup>(17)</sup>

The frequent hospitalizations, due to chronic diseases, interfere with affective bonds and sometimes cause ruptures in relationships, due to the long periods of separation. This type of event can also increase the vulnerability of adolescents with chronic disease, start a process of depression and evidence ineffective coping, changes in self-concept, and decreased self-esteem.<sup>(20-22)</sup>

In addition, the frequent hospitalizations intensify fears because the adolescents discovering new complications, due to the unpredictability of crises and the course of the disease, as each crisis can be different, reveal damage to different organs and bring new challenges to be faced. Because it is a

chronic disease, these fears will accompany the adolescents into adulthood.<sup>(23)</sup>

Several hospitalizations lead to delays in the school schedule and can provoke a feeling of inferiority.<sup>(16)</sup> The psychological and physical repercussions of SCD affect school attendance and performance, which can frustrate the prospects of professional accomplishment, affect mental health, and consequently increase the use of health services.<sup>(24,25)</sup>

Thus, “Feeling like a normal adolescent” was revealed as the strategy the adolescents used in coping with the central category seeking to be a normal teenager, despite the restrictions and discrimination the SCD imposes”. Adolescents with chronic kidney disease also reported this strategy when they observed the differences from other adolescents, trying to say that their condition was normal, as a way of affirming for themselves that their existence was common.<sup>(17)</sup> In a Jamaican study, however, adolescents with SCD felt normal the more they were involved in social activities, such as attending school, participating in sports and parties, and the less they needed hospitalizations.<sup>(26)</sup>

A study showed that adolescents with SCD valued the maintenance of “normal” life and employed strategies to minimize differences between their peers, often placing themselves in vulnerable positions, seeking to take control of their lives. In this same study, the authors found that the adolescents also compared themselves to adolescents with other chronic diseases and disabilities, considered as worse than their condition, thus sustaining a sense of “normality” and resisting the label of victims.<sup>(26)</sup>

In addition, the acceptance of their condition as having SCD revealed the adolescents’ positive coping with the adversities triggered by the disease and indicated a balance between the malaise of chronic illness and the well-being that arose when the symptoms were controlled. In this context, chronic illness was a significant problem in adolescents’ lives. To overcome it daily, the strategy they found was the affirmation of their normality, sometimes denying the disease, sometimes facing it, but minimizing its consequences, hiding what appeared to them, from their condition.<sup>(26)</sup>

By trying to be a “normal” teenager, they submitted to the therapeutic processes required in the

management of the disease to achieve quality of life, adapting to restrictions, and adopting changes in daily life. They also transgressed certain rules of self-care, such as not engaging in strenuous activities during leisure time with friends.

Also in the pursuit of normalcy, adolescents with SCD tended to hide pain to spare their parents from suffering,<sup>(27)</sup> which increased their vulnerability by delaying the care for complications. In addition, in the attempt to insert themselves among their peers, the adolescents could adopt some risky behaviors.<sup>(28)</sup>

The protection of individuality and the search for autonomy were perceived in these small ways of transgressing the care norms, because the adolescent wished to recognize himself as being able to do things by himself, to take the direction of his own life through his choices.<sup>(29)</sup> These transgressions could also express the recognition of something that bothered him and from which he sought to escape.<sup>(15)</sup>

The category “Perceiving the stigma” represented the consequences of the central category through the subcategories “Feeling ashamed of having sickle cell disease”, “Fearing discrimination because of being sick”, “Facing the stigma of sickle cell disease” and “Feeling powerless in the face of sickle cell disease”. It was evident that part of the adolescents experienced the shame of having their diagnosis revealed and this, consequently, would cause a separation from colleagues. Thus, they could suffer discrimination and social exclusion, and feel powerless because they are unable to change their condition as patients. Some reported though that when they disclosed the diagnosis of SCD, they felt more support from friends.

This aspect emerged from the experience of stigma and made the patient predict, through the feeling of shame, that a characteristic of his could be considered a reason for the estrangement of people. This feeling mobilized him to adopt the concealment of information that could make him experience distancing as a strategy.<sup>(30)</sup>

Research conducted with adults diagnosed with SCD identified that the respondents attributed the stigma experienced not to the existence of the disease itself, but to the fact that its complications prevent them from fulfilling their roles and meeting social expectations.<sup>(22)</sup> Considering this experience,

it can be inferred that the adolescents in this study adopted attitudes that allowed other members in their context to judge and frame them as “normal” and “capable” of meeting the norms and expectations of their peer group, such as attending school, participating in sports, going to parties, dating and hanging out with friends.

It is highlighted that receiving support from family, school, and friends favors the positive coping with the chronic disease and associated stigma and can reduce depression and strengthen affective family bonds.<sup>(31)</sup>

## Conclusion

The experience of adolescents with SCD is represented in the core category “Seeking to be a normal adolescent, despite the restrictions and discrimination imposed by sickle cell disease”. The causal condition of this category is the feeling of being different from other adolescents and occurs in the context of living with restrictions and having bad experiences, such as the experience of pain, constant hospitalizations, fear of death, and uncertainty about the future. Thus, the adolescent is brought to feel normal. By perceiving stigma and fearing discrimination, (s)he adopts ways to hide that (s)he has the disease. By seeking to be normal adolescents, the participants aspired to take control over their own lives, avoid breaks from the routine and meet social expectations, protecting their identity from labels and discrimination. The ways to face the stigma depended on the knowledge that family members and other people had about the disease, the support they received, and how they developed strategies to deal with these perceived differences when interacting with their peers. The concern with meeting social expectations and fitting into normal patterns increases the vulnerability of adolescents with SCD.

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## Collaborations

Santos LM, Peixinho Neta TS, Brito LS, Passos SSS, Jenerette CM, and Carvalho ESS collaborated with the study design, data analysis, and interpretation, writing of the article, relevant critical review of the intellectual content, and approval of the final version for publication.

## References

1. Soares LF, Lima EM, Silva JA, Fernandes SS, Silva KM, Lins SP, et al. Prevalência de hemoglobinas variantes em comunidades quilombolas no estado do Piauí, Brasil. *Cien Saude Colet*. 2017;22(11):3773-80.
2. Piccin A, Murphy C, Eakins E, Rondinelli MB, Daves M, Vecchiato C, et al. Insight into the complex pathophysiology of sickle cell anaemia and possible treatment. *Eur J Haematol*. 2019;102(4):319-30.
3. Mastandréa EB, Lucchesi F, Kitayama MM, Figueiredo MS, Citero VA. The relationship between genotype, psychiatric symptoms and quality of life in adult patients with sickle cell disease in São Paulo, Brazil: a cross-sectional study. *São Paulo Med J*. 2015;133(5):421-7.
4. Reisinho MC, Gomes B. O adolescente com fibrose cística: crescer na diferença. *Rev Port Enferm Saúde Mental*. 2016;(Esp 3):85-94.
5. Santos GS, Tavares CM, Aguiar RC, Queiroz AB, Ferreira RE, Pereira CS. Pursuing online health information: coping strategies of adolescents with chronic diseases. *Rev Port Enferm Saúde Mental*. 2016;(Spe 4):33-8.
6. Smith SK, Johnston J, Rutherford C, Hollowell R, Tanabe P. Identifying social-behavioral health needs of adults with sickle cell disease in the emergency department. *J Emerg Nurs*. 2017;43(5):444-50.
7. Freitas SL, Ivo ML, Figueiredo MS, Gerck MA, Nunes CB, Monteiro FF. Quality of life in adults with sickle cell disease: an integrative review of the literature. *Rev Bras Enferm*. 2018;71(1):195-205.
8. Abreu IS, Nascimento LC, Lima RA, Santos CB. Crianças e adolescentes com insuficiência renal em hemodiálise: percepção dos profissionais. *Rev Bras Enferm*. 2015;68(6):1020-6.
9. Silveira A, Neves ET. Daily care of adolescents with special health care needs. *Acta Paul Enferm*. 2019;32(3):327-33.
10. Paris de Souza I, Bellato R, Santos de Araújo LF, Barros de Almeida KB. Adolescer e adoecer na perspectiva de jovem e família. *Cien Enferm*. 2016;22(3):61-75.

11. Charon JM. Symbolic interactionism: an introduction, an interpretation, an integration. New Jersey (US): Prentice-Hall; 2010.p.22-33.
12. Strauss A, Corbin J. Pesquisa qualitativa: técnicas e procedimentos para o desenvolvimento de teoria fundamentada. 2a ed. Porto Alegre: Artmed; 2008. p.65-209.
13. Brasil. Ministério da Justiça. Lei 8.069, de 13 de Julho de 1990. Dispõe sobre o Estatuto da Criança e do Adolescente e dá outras providências. Brasília (DF): Ministério da Justiça; 1990 [citado 2021 Maio 22]. Disponível em: [http://www.planalto.gov.br/ccivil\\_03/leis/l8069.htm](http://www.planalto.gov.br/ccivil_03/leis/l8069.htm)
14. Coutinho MP, Serafim RC, Araújo LS. A aplicabilidade de desenho-estória com tema no campo da pesquisa. In: Coutinho MP, Saraiva ER. Métodos de pesquisa em psicologia social: perspectivas qualitativas e quantitativas. João Pessoa (PB): Editora Universitária; 2011. p. 205-50.
15. Souza MA, Melo LL. Being adolescent with chronic renal failure: a view through existential phenomenology. *Esc Anna Nery*. 2018;22(2):e20170368.
16. Kelly AD, Egan AM, Reiter-Purtil J, Gerhardt CA, Vannatta K, Noll RB. A controlled study of internalizing symptoms in older adolescents with sickle cell disease. *Pediatr Blood Cancer*. 2015;62(4):637-42.
17. Marques T, Vidal SA, Braz AF, Teixeira ML. Clinical and care profiles of children and adolescents with Sickle Cell Disease in the Brazilian Northeast region. *Rev Bras Saúde Mater Infant*. 2019;9(4):881-8.
18. Cecilio SG, Pereira SA, Pinto VS, Torres HC. Barriers experienced in self-care practice by young people with sickle cell disease. *Hematol Transfus Cell Ther*. 2018;40(3):207-12.
19. Alveno RA, Miranda CV, Passone CG, Waetge AR, Hojo ES, Farhat SC, et al. Pediatric chronic patients at outpatient clinics: a study in a Latin American University Hospital. *J Pediatr*. 2018;94(5):539-45.
20. Graves JK, Hodge C, Jacob E. Depression, anxiety, and quality of life in children and adolescents with sickle cell disease. *Pediatr Nurs* 2016;42(3):113-9,144. Review.
21. Lukoo RN, Ngiyulu RM, Mananga GL, Gini-Ehungu JL, Ekulu PM, Tshibassu PM, et al. Depression in children suffering from sickle cell anemia. *J Pediatr Hematol Oncol*. 2015;37(1):20-4.
22. Ola BA, Yates SJ, Dyson SM. Living with sickle cell disease and depression in Lagos, Nigeria: a mixed methods study. *Soc Sci Med*. 2016;161:27-36.
23. Coleman B, Ellis-Caird H, McGowan J, Benjamin MJ. How sickle cell disease patients experience, understand and explain their pain: an interpretative phenomenological analysis study. *Br J Health Psychol*. 2016;21(1):190-203.
24. Prussien KV, DeBaun MR, Yarboi J, Bemis H, McNally C, Williams E, et al. Cognitive function, coping, and depressive symptoms in children and adolescents with sickle cell disease. *J Pediatr Psychol*. 2018;43(5):543-51.
25. Foster N, Ellis M. Sickle cell anaemia and the experiences of young people living with the condition. *Nurs Child Young People*. 2018;30(3):36-43.
26. Forrester AB, Barton-Gooden A, Pitter C, Lindo JL. The lived experiences of adolescents with sickle cell disease in Kingston, Jamaica. *Int J Qual Stud Health Well-being*. 2015;10:28104.
27. Atoui M, Badr LK, Brand TD, Khoury R, Shahine R, Abboud M. The daily experiences of adolescents in Lebanon with sickle cell disease. *J Pediatr Health Care*. 2015;29(5):424-34.
28. Asnani MR, Bhatt K, Younger N, McFarlane S, Francis D, Gordon-Strachan G, et al. Risky behaviours of Jamaican adolescents with sickle cell disease. *Hematology*. 2014;19(7):373-9.
29. Labore N, Mawn B, Dixon J, Andemariam B. Exploring transition to self-management within the culture of sickle cell disease. *J Transcult Nurs*. 2017;28(1):70-8.
30. Lacerda FK, Carvalho ES, Araújo EM, Miranda NB, Dias AL, Almeida TA. Women with sickle anemia living with leg ulcers and pain. *J Nurs UFPE On Line*. 2014;8(7):2054-60.
31. Harmelen AL, Gibson JL, Clair MC, Owens M, Brodbeck J, Dunn V, et al. Friendships and family support reduce subsequent depressive symptoms in at-risk adolescents. *PLoS One*. 2016;11(5):e0153715.