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Experiences and strategies of people with sickle cell disease in the Federal District: the biographical rupture

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Abstract *This is a study on sickle cell disease, a chronic illness that affects many Brazilians, that aims to understand and analyze how people address the adversities arising from the diagnosis and the biographical rupture. The description of people's experiences and strategies conjures a picture that expresses the respondents' habitus, built in a dialectical relationship with the vulnerability determined by the disease. We adopted a qualitative approach and focused interviews as proposed by Merton, combined with the snowball technique, applied to groups related to sickle cell disease on social networks. Seven participants were selected because they were privileged informants with the disease, were over eighteen, lived in the Federal District, and were non-exclusive users of the Unified Health System. The interview material was categorized from the focal groups employed. The results indicated the following categories: biographical rupture, experience and coping strategies, and healthcare. It is necessary to sensitize professionals and the population about the challenging living conditions of people with sickle cell disease and the consolidation of public policies and care networks to accommodate this population.*

Key words *Sickle cell disease, Chronic disease, Experience, Social vulnerability*

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Introduction

Sickle cell diseases are chronic genetic diseases, and several discussions exist about the etiology and genetic mutations from which the diseases are derived[1]. In this work, regarding the biomedical definitions of the disease, we will use the term sickle cell disease because, per researcher Silva, the term encompasses all other disorders caused by the same genetic mutation besides sickle cell anemia[2].

Contrary to popular belief, sickle cell disease is not a rare disease. Modell et al.[3] argue that it is part of a large group called "hemoglobin disorders". In sickle cell disorders, sickled red blood cells block small blood vessels and cause anemia, functional asplenia, severe pain episodes, and residual organ damage. Since then, several studies have been conducted to describe the pathological aspects and physiology of sickle cell disease; that is, the best ways to understand the association between the disease and the needs of the people receiving care are being studied to this day[4].

According to Model et al.[3], such disorders are found in 71% of the 229 countries in the world, and 5.2% of the world population carry the responsible gene. Furthermore, hemoglobin S is responsible for 40% of carriers and causes more than 80% of the comorbidities of people with this disease. Therefore, it is estimated that around 276 thousand children are born with sickle cell disease per year in the world, of which 3.5 thousand would be born in Brazil[5].

The disease is diagnosed early through Neonatal Screening, popularly known as the "heel prick test", a test run from the third to the sixth day of life[6]. It is worth remembering that a child's right to take the test anywhere in Brazil[7] became effective only in 2001. Before this standardization, the disease could be diagnosed from the first six months of life through the child's symptoms and clinical complications, but the diagnosis could take time[8].

Nascimento et al.[9] affirm that two age groups are most affected: 0-4 years and 15-19 years. The authors point out that death is a possibility in adolescence, even with different care needs and after overcoming all the problems during childhood arising from sickle cell disease. It is critical to building full personal awareness and transitioning to adulthood. They point out that "elucidating this problem will be essential to readjust attention at this stage of life, which is full of challenges and uncertainties and deteriorated by SCD"[9] (p. 7).

People with sickle cell disease evidence several clinical symptoms, such as pain attacks, fevers, sudden spleen enlargement or abdominal distension, chest pain, cough, visual impairment, prostration, difficulty breathing, painful penile erection, nausea, vomiting, jaundice, urological changes (convulsions, lethargy, muscle weakness, behavioral change), inability to drink fluids and dehydration signs. Therefore, we can see that the health complications of these people do not allow them to have full conditions and quality of life, requiring public and social policies to protect and allow them to participate as citizens[10]. The only "cure" described is bone marrow transplantation, a proposal still being studied in academic circles, and the long-term perspective must be considered even with the benefits of bone marrow transplantation in the short term; besides that, treatment is still restricted to specific groups[11].

Theoretical framework

The global and Brazilian demographic transition has led us to the continuous growth of chronic illnesses typical of modern societies[12]. More than acute diseases, these diverge from the Natural History of Disease model as they do not have a cure or total rehabilitation.

Our motivation for the research was to elucidate how sickle cell disease causes suffering, loss, and incapacity in routine activities, which we call biographical rupture. This rupture can occur at different stages of someone's biography, introducing new experiences, such as the need for medication and care spaces, and economic, religious, and social complications endured by people with this disease[13].

Michael Bury considers that the disease is related to social and economic issues that interfere with the experiences of people in the chronic illness process, generally catalyzed by the diagnosis and described by Bury[14,15] as biographical rupture, when individuals begin to experience the loss of their life routine and need to reinvent or develop new practices to continue their "self" and meet new responsibilities. Chronic illness breaks the structures with which the individual is accustomed to living and knowing, leading to facing new realities that involve pain, suffering, and the possibility of death, as found differently in other studies[16-18].

Given the importance of the topic, this research aimed to elucidate how people manage and administer their lives with the disease and

its symptoms – their experiences, based on the biographical rupture caused by sickle cell disease. We analyzed the strategies people adopt and incorporate into their habitus, or rather, how these people's habitus manifests itself in the daily practices of their treatment and when relating to their bodies. To do so, we employed the concepts that we will discuss below.

Pierre Bourdieu's concept of habitus[19] refers to historical learning through the secondary family interaction at school and tertiary in forming professional habitus. The habitus transcends individuality, as it incorporates learning and acting in situations in the social agent's social fields, creating marks in thinking and acting according to the group, time, and people with whom one lives. Montagner and Montagner[17] (p. 197) affirm that:

Habitus indicates a system of schemes of perception, values, judgments, appreciation, and action inscribed in the body, incorporated by experiences like "dead branches" that still belong to the tree and are part of its structure.

Knowledge of the expressions of the habitus of people or groups through practices for coping with chronic illness will highlight unknown aspects of the relationship between individuals and the illness and guarantee the development of new care proposals for this population through actual data that understands the individual demands. In coping with the disease, people employ practical and pragmatic strategies based on their life experience or creativity in the moment of need. These strategies depend on the resources available to the individual – which could be called "capital" in Bourdieu's theory – and are intrinsically linked to the group habitus.

The agents' expression and realization occur in social fields in which the presence of the disease determines the emergence of different vulnerabilities in a group[17]. The vulnerable situation of people with sickle cell disease, arising from social structures, leads them to employ daily strategies to cope with their condition. These strategies are part of the habitus of individuals and their groups, as this habitus provides the means and tools of action that are continually put into play. In a structured and structuring way, these means in the habitus can be understood as economic, social, cultural, symbolic and other capitals[20]. This historical conformation of vulnerability leads individuals to more or less effective practices within their living conditions and a given social field.

Montagner and Montagner[21] describe structural or genetic vulnerability as a defined situation in a historical moment of concrete origin derived from a particular social field, recognized in social and relational terms by groups, which involve more than only the individual and determine their group. Genetic vulnerability in social structures reaches all group members in question through the conditions of culture, politics, and economics, that is, all social fields. Although in indirect or diffracted forms by social fields and class determination, the structural vulnerability – of the group – will lead all these individuals to be devalued as public figures and citizens, which can lead to an alienation of their rights as humans and citizens.

The central objective of this article is to understand and analyze how people address biographical rupture and adversities from their vulnerability by describing the experiences and strategies of people with sickle cell disease.

Methods

The qualitative approach was used in this study. We adopted the focused interview, described by Merton et al., which requires prior knowledge about the subject or event addressed and that interviewers and respondents must have a direct relationship with the theme[22]. In this technique, the focus cores of the interview script are established beforehand from the consulted bibliography and the researcher's experience. These cores direct the interviews and delimit the subject. The semi-structured interview was divided into four focus cores of questions: life history, sickle cell disease, experiences, and specific coping strategies.

This research was submitted to and approved by the Research Ethics Committee of the University of Brasilia, Ceilândia Faculty, File nº 25406619.2.0000.8093.

The inclusion criteria adopted in the study were to have been diagnosed with the disease, exclusive use of the Unified Health System (SUS) in the Federal District, to be 18 or over, to live in the Federal District (DF) or RIDE (Integrated Region of Economic Development) of the DF and be part of some social or political organization whose central objective is sickle cell disease.

The participants were selected as privileged informants because they were people with social-political action, either in organized groups in patients' associations or with a relationship with politicians and public agents such as doctors, managers, and academics.

We adopted the snowball technique based on gathering individuals with a common feature in the sample, regardless of their population group. The first participants were contacted in communities (social pages and networking groups) related to sickle cell disease and addressed directly, explaining the research. We asked them to indicate people with the same characteristics defined for the study. This process was repeated until a "snowball" of participants was achieved and the desired sample and research saturation point was reached[23]. The interviews were conducted virtually using Google Meet, Zoom, and WhatsApp.

The study was conducted from May to June 2020 and was attended by seven people. Fictitious names were based on personalities of Brazilian popular music (MPB), namely Elis, Clara, Clementina, Marisa, Geraldo Vandré, Adoniran, and Nilze to preserve their identities. We gathered findings around three categories with these interviews: biographical rupture, coping strategies, and healthcare.

Results and discussion

Socioeconomic data

Two men and five women were interviewed. Participants declared themselves as Black (3), brown (2), white (1), and one did not respond. Four respondents considered themselves Roman Catholic; one was non-practicing Roman Catholic, one was evangelical, one had no religion, and one did not respond. Five were married, and two were single. Finally, four had a private health plan.

Participants resort to SUS in times of crisis, especially the reference centers, even if they have health plans with varying coverage levels (generally more straightforward plans) and receive medications through the SUS high-cost pharmacy. Health plans are used to treat comorbidities and perform tests with faster results. Some participants reported using health plans for mental health treatments, such as psychologists and psychiatrists.

Most of the sample stems from the Black population, composed of Blacks and browns. We highlight the link between sickle cell disease and the racial dimension. As Calvo[24] points out, the concepts related to racial aspects are defined by history and culture over the ages. The relationship between medicine and common sense does not escape this rule. The concept of race, understood by medicine and common sense, has undergone historical and cultural changes throughout the sickle cell disease study's history. Changes we find in today's society regarding understanding the sickle cell disease related to Blacks and browns originate from the Black population's movements to revisit politically and ideologically the structures of service, reception, and care of individuals per their ethnic origin.

The participants' age bracket was 33-54 years. Three participants claimed to be retired; one was a nursing technician, two were civil servants, and one was a psychologist. Relevant data found was that the retirees had elementary or high school levels. In contrast, those with technical or higher education continued to work in their areas after they made the necessary adaptations. The possible explanation is the disease and its complications, which prevent work with more rigid schedules or physical performance. This would be facilitated in the more "liberal" professions with greater flexibility in schedules and time availability.

These informants were over 33 and had undergone significant hospitalization periods, tests, transfusions, and surgeries, as there were no other drugs and curative possibilities thirty years ago. While not a clinical practice guide, the document issued by the National Committee for the Incorporation of Technologies in the SUS (CONITEC)[25] on the impacts of new and emerging drug technologies and SUS for the treatment of sickle cell disease unveiled the number of new drugs incorporated in recent years. These medications increase the number of life expectancy and positively affect the quality of life, altering perception and experience with the effects of this disease.

Biographical rupture

The diagnosis is generally the initial milestone in the construction of the individual's habitus with the disease. From then on, the social agent starts to internalize and become aware of his situation of chronic sickness and the changes in his daily life. This is when we can consider biographical rupture[15]. Biographical rupture can be a gradual process, undergoing increasing in-depth degrees.

Sickle cell disease is detected through the well-known "foot test", which became mandatory in 1992 with Ordinance n° 22 of the Ministry of Health that established the Early Diagnosis of Congenital Hypothyroidism and Phenylketon-

uria Program. In 2001, neonatal screening was expanded to cover 100% of births. However, Mallmann et al.[26] point out that the test has a positive relationship with the region of the country, with the highest likelihood of performing in the Southeast and South, then Midwest and Northeast compared to the North, with a higher likelihood among those with health plans, the highest per capita household income, and white. Finally, misinformation or ignorance about the importance of the test is another inequality pointed out[27].

Considering our informants' age, not all received a timely diagnosis. They said that the mothers had to see several specialists to obtain a diagnosis. There were even accusations of child mistreatment, as they had swelling on their hands and feet due to continuing crying in pain. The first rupture should have occurred at school, where differences began to be observed.

Clementina, for example, reported issues typical of biographical rupture, as being Catholic, she had difficulty attending mass and, besides her chronic illness, had complications resulting from radiotherapy treatment for pelvic cancer. Such limitations prevented her from carrying out activities that she considers essential for her life, requiring her to reinvent her daily actions and routine. She expressed that the impossibility of kneeling to say her prayers often made her feel bad in front of her fellow believers. Being unable to express her faith had a much more significant impact than being unable to care for herself.

A fundamental point reported by several respondents was the limited choice of their professions due to personal limitations (regardless of personal interests) and leaving formal studies. Only attending classes or working for health reasons leads to employers' and teachers' understanding. This rupture condition is reinforced in Adoniran's statement:

It's complicated having to fight the disease every day and prove your competence so as not to be fired from your job or be able to keep up with your schoolmates. Not everyone can do it. We already fight too much for our lives.

The frequent demand for leaves of absence and medical certificates is interpreted as laziness and inability to work or study. Even if it is to get medication, the time required for care in Health Units varies with the demand, but all participants reported that emergency care takes time if it is provided in emergency rooms or Emergency Care Units. Adoniran mentions that the service takes one to two hours, even when he receives the orange bracelet indicating the severity of the condition.

Routine appointments with the hematologist are relatively straightforward. However, rescheduling the appointment is complicated if something prevents attendance on a specific day and time. Geraldo Vandré reports that he periodically visits Health Units to assess his health after a liver transplant. He already underwent a bone marrow transplant and always prioritizes scheduling dates, as he knows that he will leave the service flow if he misses them.

Another example regarding the biographical rupture is that vacations are no longer pleasant and bring new concerns. Someone with sickle cell disease should consider where to get medical care if they have a crisis. This is what Adoniran narrated:

When you go on a trip, you just choose the clothes you will wear, right? In my case, I also have to have a [hospital] place of care, medications, and other things that I need to prepare, or else we cannot travel. Moreover, even so, I know I will have complications when arriving at the location.

Coping strategies

As a chronic illness, sickle cell disease requires several strategies to allow the sick person to experience everyday situations and social demands without significant losses. To do this, people require a health monitoring routine, as many factors can lead to crises. How do these people achieve this support, especially in the SUS? Some of these strategies are presented in the following paragraphs.

Clara reports that she had to hide her chronic illness to obtain medical insurance, given that health insurance plans do not authorize membership due to possible increased costs with treating complications. Adoniran, who does not have medical insurance, also states that it is hard to get a health insurance plan if you have a chronic illness and, therefore, he has never even tried to sign up for a membership contract. Thus, it is clear that people with sickle cell disease tend to be treated only by the SUS or struggle to obtain health care. Thus, the possibility of obtaining adequate treatment to remain healthy and without crises has been removed from their lives: the private sector does not want to assume this responsibility and cost and has difficulties meeting this demand.

These people's health strategies become even more complex when considering obtaining med-

ications through high-cost pharmacies. All informants reported difficulties accessing at least one of the medications they use. In cases of lack of access, it is necessary to purchase the medicine. Therefore, another widespread strategy for patients is to ask groups of people with sickle cell disease on social media if anyone has medicines left. This strategy of bartering medicines is prevalent. Nilze, for example, needs to buy each of her medications, except for sickle cell disease medications, to obtain prescribed medication for depression and panic syndrome. This expense is very significant for her financial condition, which is why she accepts help and barters to get the medications. These strategies can have several consequences, from taking medication with the wrong dosage to taking medications without a prescription, and both cases can trigger other health problems.

Regarding information and "appointments", exchanging information between participants on social networks about sickle cell disease is one of the strategies used. In these groups, information is provided on where medicines are dispensed, recommendations for hospitals and doctors, and each situation concerning their health and procedures performed. Groups feel safe when they have people who face the same problems and provide physical, moral, and even religious support.

Mutual help between group participants on social networks positively affects adherence to medication treatment, as there is an exchange of information about side effects, relief practices, and ways to obtain medication and medical care. Some respondents pointed out that a doctor provided emergency care and helped with clinical, political, and legal issues in one of the groups. However, this does not prevent self-medication, encouraged by some people in the group, as Adoniran pointed out:

[...] it is always necessary to be careful with information. Some overvalue medicines that still need confirmation, while others give tips on home remedies. I've already been involved in this, and I can assure you that we have to be careful, as I've already been a victim of this misinformation, and I always fight when I see something like this in groups.

Another strategy people use is judicialization. Research participants reported that a quicker way is often to take legal action instead of seeking medication or hospitalization through the usual means of services, as this is sometimes encouraged by service professionals. Sometimes, while a relative accompanies the sick person to the hospital or pharmacy, another person shows up at the Public Defender's Office.

Healthcare

Two care types meet the health needs of this vulnerable population: emergency and urgent care, on the one hand, and routine appointments and interventions, on the other. All of our informants used it. However, there is a caveat. This study was not about evaluating care but analyzing the strategies used by participants to receive care in the most different healthcare settings. In general, respondents commented on the difficulty in scheduling more complex tests. Therefore, they try to run tests in private clinics, increasing costs. However, doing so speeds up the therapy progress through the SUS.

For those interviewed, accessing a trusted doctor is essential. They reported preferring to wait longer to receive care from specific professionals, as they know their ethical commitment and involvement with people. They emphasized the professionals' social nature, sometimes to the detriment of their knowledge, even considering that they are very qualified and understand a lot about the disease since, in general, their needs are resolved in a short time. All informants praised the role of their doctors in providing swift, efficient, and attentive care.

The search for care is related to extreme pain, priapism, fever, and shortness of breath, and generally to confirm or not any sickle cell disease complication. A frequently reported fear is that of receiving treatment for iron deficiency anemia, and, according to informants, this has already happened, and the result was death from septicemia. Therefore, they make a point, even at the height of their pain, to make it clear that it is "sickle cell disease" and not "sickle cell anemia".

Another severe problem that can sometimes prevent care is being confused with drug users in a withdrawal crisis. This is because they already know and demand the medications and dosages that should be administered, but also because of their physical appearance. People with sickle cell disease report that they often arrive at the health service shaking due to pain, their eyes may be yellow due to the anemia effect, they are in a state of priapism, and with other symptoms that can confuse professionals. Patients tend to suffer discrimination, given the racial dimension, combined with these stigmatizing marks.

A public policy that helped them a lot was creating a card containing all their data, making

it easier to identify the disease and who to turn to in case of doubts. For example, Adoniran says that he felt nausea, lethargy, and hearing hallucinations. He believed that these were adverse effects related to pain medication, and he did not use any medication or alcoholic beverages. Therefore, he is not in the group of drug users: "I'm not prejudiced, but I don't use drugs, and I'm sure that no one who has sickle cell disease does." Geraldo Vandré corroborates Adoniran's version, emphasizing that not taking medication is a miracle for them: "I couldn't believe it when I realized that I would no longer have to use medication [opioids]. I felt free".

Necrosis of the femoral head and other joints requires surgery and frequent physical therapy. Besides the difficulties of necrosis itself, such as locomotion and mobility hardships, we emphasize sexuality constraints, as they hinder the very sexual intercourse act. We should mention that men with a priapism crisis caused by the disease tend to be misunderstood or confused in many situations in the services. This leads them to seek help only when the situation becomes severe, which can lead to more drastic treatment measures[10].

Nilze stated that she had osteonecrosis of the femoral head and knees and needed two surgical interventions to regain functionality. She also mentioned that the hospital where she was being monitored refused to perform the last surgery due to the high risk and did not offer follow-up with a physiotherapist and occupational therapist. The second surgery was performed in a private establishment, so she covered all these expenses.

Regarding reproductive life, the information we received from participants is that some health professionals tend to be discouraged and sometimes give misleading guidance about genetic counseling for planning and deciding to have a child. Others informed us that the decision to have children was not discouraged by health professionals, but the fear of having children with the disease made them give up the idea.

On the other hand, with the advent of bone marrow transplantation and new genetic treatment techniques for sickle cell disease, parents are encouraged to have other children to obtain material from the umbilical cord. There was an account that this has already happened, although there is a risk of having another child with the disease, as it would be worth trying:

I know someone who had another child in an attempt to have a transplant. However, it was not possible as the child was born with even greater complications. Nothing is certain (Adoniran).

Blood transfusions act as a prophylactic measure against the emergence of complications such as pain crises and strokes. In some cases, it is the only measure indicated to prevent pain crises and complications[28]. All informants had blood transfusions and said that, in this case, they received the necessary care, and this happened because it was an action the health system classified as an emergency.

Adoniran reported that he needs appointments every twenty-one days in which the need for blood transfusions is assessed. Adoniran also needed medical care due to the severe pain he was feeling. However, he has recently adopted the strategy of receiving care from the Mobile Emergency Care Service (SAMU). Calling in this service is because the patient already receives pain medication inside the ambulance and, if necessary, professionals can arrange hospitalization more easily. He started using this method after they called SAMU to assist him first, and he saw that it was a good possibility.

Marisa, in turn, required more specific care due to leg ulcers, a stomach ulcer, and hepatitis caused by medication. According to Dienstag[29], Liver diseases caused by medications, such as hepatitis, described by Marisa, are the most common among acute liver diseases. One of the questions the group asks is how one knows if the signs and symptoms are due to sickle cell disease or if they are events that have nothing to do with the disease, thus requiring other care. The informants said that the disease explains all they feel.

In all cases and situations reported, we can witness a high dependence on health services in primary care and reference centers. This dependence makes them known since childhood in specific places, which makes it easier for professionals to understand the protocols well, and they already know what to do, the medication dosages, the need to explain the disease itself again, all this due to the bond established between patients/professionals. However, it does not mean they will have priority, perform tests, or obtain medications more easily.

Final considerations

Although the findings cannot be directly generalized, the different aspects presented by the participants when reporting their experiences reinforce two central arguments involving chron-

ic illnesses. The **first aspect** is that the disease affects people from different social groups and realities, and, therefore, the illness process is diverse and individualized, even though it follows specific patterns of demands and occurrences that make people use their strategies or in groups to mitigate and overcome hardships.

Psychological and psychiatric support should be an available and frequent practice in cases of chronic illnesses, especially those in which people feel constant pain and during periods of more intense pain crises. Legal support would serve in cases of complications of the disease and its consequences, which could lead to retirement and pension, among other demands.

The analysis of the biographical rupture clearly showed us that life strategies must be continually updated and re-elaborated after the diagnosis[30]. Among the people researched, many went through discovering the disease, as they were born before the heel prick test was introduced, and the rupture unfolded well after birth[15]. The disease is a factor that affects people from different social classes and contexts, making those affected a diverse group with very heterogeneous experiences and defense mechanisms despite the same group's structural vulnerability. Those who had an early diagnosis went through a biographical rupture right at the beginning of their upbringing as individuals; others went through a continuous process that sometimes reached adolescence, with acute episodes and stages of relative adaptation to the illness, until complete rupture, as pointed out by Bury[15,31]. In this context, *habitus* makes people deal uniquely with the adversities throughout their life trajectories, even while building social networks to support or share their strategies.

The **second aspect** is related to the difficulties and role of the Unified Health System. Equity is one of SUS pillars and must be applied directly to reduce the hardships and barriers in health care. There should be a dialogue between different spheres of care so that people are assisted, and their difficulties respected.

Political institutions, when requested, meet the demands of organized civil society. We have found that Brazilian states with an official representation or association of people with sickle cell disease obtain the best results for those affected. It is necessary to sensitize health professionals and those linked to the disease about adversities and the consolidation of robust and ubiquitous policies and care networks.

We want to state that one of our informants passed away before we concluded our study, and we dedicate this research to him.

Collaborations

JL Costa: designed and conducted the research, wrote the initial version of the article and participated in the final review. MI Montagner and MA Montagner: designed the research, guided work, wrote the article, and performed the final review. SMC Alves and MC Delduque: participated in the final review.

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