

Research Article**Sickle cell disease and projection into future in African patients living in France, born in Africa and those born in France from African Parents****Hassan Njifon Nsangou**

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Abstract

Introduction: Sickle cell disease is the most common genetic disease in the world. In France, there are patients born in this country and those born in their country of origin. This work, which raises the problem of the experience of the disease by these two categories of patients, questions their projection into the future with a view to improving their care.

Material and method: 20 subjects from sub-Saharan Africa (12 women and 8 men) and residing in France for at least two months, aged 19 to 71, including 10 born in France and 10 born in Africa participated in this research. The data was collected via clinical interviews and the quality of future projection questionnaire carried out in a Parisian hospital.

Results: The analysis of the data highlights the predominance of magico-religious representations among subjects born in Africa who plan for their future with little assurance and security, unlike those born in France. It shows the advantage that patients born and cared for in France have compared to those who were born and cared for in Africa before their arrival in France. In addition, subjects born in Africa show a tendency to give meaning to sickle cell disease by relying more on cultural representations without however turning away from biomedical knowledge related to this disease. An opposite trend is observed among subjects born in France concerning the meaning given to illness.

Conclusion: The research highlights the issue of the cultural, social and medical environment on the process of subjectivation of sickle cell disease by adults affected and further shows, following other research, the need for interdisciplinary support for patients in an ethno psychiatric device.

Keywords: Sickle cell disease; Culture; Projection into future; Africa; France.

Introduction

Sickle cell disease (SCD) is a serious chronic disease that manifests itself from early childhood with intense pain, chronic anemia, infections to which can be added acute and chronic complications affecting the brain, heart, lungs, and kidneys, among others. All organs can be affected. A genetic disease, it is transmitted to the child by both parents, each carrying the S gene for sickle cell disease. SCD is the most widespread genetic disease in the world (1) with around 150 million healthy carriers of the AS trait, 2.3% worldwide (2). It mainly affects the black population and is very common in sub-Saharan Africa, in the Mediterranean basin, in the Maghreb and in the Middle East, in India, in Brazil, in the United States and in practically all countries with a population originating from Sub-Saharan Africa (3).

In France, SCD is a rare disease and the most common genetic disease affecting 15,000 to 20,000 patients, most of whom live in Ile de France and in the overseas departments and territories (4). SCD crises are relieved, depending on the case, by hydroxyurea, blood transfusions, bleeding and analgesics. Bone marrow transplantation is currently the treatment that seems to be very effective against severe sickle cell crises (5), promising gene therapy avenues are being studied (5).

Psychological research has focused on both the experience of people with SCD and their families (6) both in hospitals and at patients' homes, paying particular attention to the culture of the subjects and their experience with this disease. During clinical consultations in hospitals in Paris, we saw a difference emerge in the way patients born in France to African parents talk about their illness and their life plan compared to those born in Africa. For patients born in France, explaining and elaborating on sickle cell disease goes without saying: "I talk about it when I want, to whom I want". For patients born in Africa: "I hurt when I talk about it, I am ashamed to talk about it", talking about it is a source of suffering. These different attitudes raise questions:

- Does the weight of cultural representations influence the psychological experience of subjects with sickle cell disease?

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- Does the representation that a subject has of his disease influence the way in which he invests his future?

- Is there a difference in the representation and experience of sickle cell disease among patients depending on their place of birth (in Africa or in France)?

This article aims to understand the experience of sickle cell disease in adult patients depending on whether they were born in France or African parents and benefit from medical monitoring from birth or in Africa and having lived there for a few years with medical monitoring. Understanding the experiences of these patients can enable professionals to improve care by taking into account the psychosocial specificities of patients. This research takes a cross-section of the experience of the disease and the projection on the future in two groups of patients met in a Parisian hospital center, patients born in France and patients born in a sub-Saharan African country, all followed in France. This cross-examination is envisaged to understand, if there are any, the specificities of the experience of sickle cell disease in connection with the place of birth of the patients, with a view to adapting care to these specificities, obviously taking into account the particularity of each case.

Theoretical Considerations

SCD is a chronic and lethal disease, supporting cultural representations.

Management of sickle cell disease in sub-Saharan Africa

In sub-Saharan African countries, care for people with SCD remains limited, due to a lack of financial means, technical platforms and political will on the part of certain governments who do not make it their priority (7). Very frequently, parents are victims of stigmatization by the population and try to hide the disease from those around them, especially since they are perceived as witches or cursed after the successive deaths of their young children (8). Because of the lack of knowledge and the incurable aspect of SCD, families seek solutions to this evil by resorting to traditional medicine.

In a survey carried out in Côte d'Ivoire (7), SCD is thought of as a disease that "lowers the blood, which kills children", a disease that cannot be cured; an incurable disease. A Malian traditional practitioner specifies that the person with SCD "stays in bed, they cannot work... it's a pity...the person grows poorly and remains stunted" (9). Sickle cell disease is therefore a handicap for the sick person and their family. Although today people are aware of the medical progress that makes it possible to contain and relieve the painful crises linked to SCD, this disease remains associated with the impotence of medicine, even that of "whites" (9). In addition, the loss of several children from the same woman feeds suspicions and diffuse beliefs (9).

SCD is perceived as "a disease of weak blood, tainted blood" which results from a poor diet or a transgression of an ancestral norm by the mother of the child affected during her pregnancy (10). This representation allows the population to give meaning to the fact that some children of the same sibling escape sickle cell disease (11). The random nature of the hereditary transmission of this disease therefore reinforces this representation (11).

Traditional sickle cell therapy

In the African conception, the origin of evil is always exteriorized (ancestor, jinn, God, Spirit, etc.). The diagnosis of a disease and the

management of the sick person are determined by the search for what may have been at stake and/or the ancestral law which has been transgressed. The disease is in fact invested by the traditional healer as an evil which says, like a messenger, an attack on the social and relational order. Illness is, in this sense, a message addressed to the patient's social and family group. The disease is in fact more that of the group than of the patient himself (10).

In connection with the invisible world, the traditional therapist identifies the origin and the meaning of the symptoms which can be a punishment, a warning, an attack or revenge and then proposes a remedy in order to restore balance in the community (12). The traditional healer can then address a request for forgiveness to the ancestors by the relatives of the patient (13) via, for example, animal sacrifices, offerings to the poor in a context of globalization.

Specificities of care in Africa

The contemporary African subject is situated between tradition and modernity. His relationship with spirits and occult forces is sometimes still very significant and more sensitive in the countryside far from specialized care centers, generally located in urban areas. Faced with the disease, he seeks help first in traditional treatments (plants, massages, incisions to penetrate the remedy, scarifications...), in visits to charlatans, to spiritual masters. Tradition is neither past nor outdated for the African (11) who relies on knowledge from his tradition and on that from modernity to give meaning to the situations he experiences (14). The frequent and painful crises of children with sickle cell disease force parents living in rural Africa to go to city to consult a doctor whose care is often expensive and not reimbursed by social security (6).

In most sub-Saharan African countries, there is still no systematic screening for SCD. The discovery of the disease in children is made by chance, for example during a consultation for repeated painful crises, frequent infections or on the occasion of anemia (5). In Africa, when there are prevention and screening campaigns, the public often tends to remain reluctant to biomedical tests, despite the severity of the symptoms observed.

Numerous awareness campaigns are undertaken with the populations to change their view of SCD, which remains an incurable and costly disease for families (5). This leads some patients and their families to immigrate to Western countries in search of better care.

Specificities of the management of sickle cell disease in France

In France, SCD is a rare disease, the treatment of which is 100% reimbursed by social security as specified above. This care begins with neonatal screening, the program of which was set up in 1995. Any three-day-old child in the so-called "at-risk" population is subjected to it (15). In the event of a positive diagnosis, during a genetic counseling consultation, the doctor explains the disease to the parents, its mode of transmission and specifies the protocol for taking care of the child in a specialized centre.

SCD is not considered an incurable disease there, due to the availability of gene therapy and bone marrow transplantation; considered effective treatments for sickle cell disease (5).

Some patients born in Africa and having fewer seizures only discover their disease once in France due to systematic screening for sickle cell disease in most French hospitals where people from Africa are tar-

get populations for screening (7). Families who settle in France, unlike those in their countries of origin in sub-Saharan Africa, benefit from 100% healthcare reimbursement. SCD is in fact a rare disease and a long-term condition in France (4). These families develop less resistance to blood transfusions; which is not often the case in their country of origin where they often refuse these transfusions for fear of contamination of their child by blood probably infected with diseases on the one hand and for fear of possession of the sick person's blood by a stranger through blood transfused on the other hand (16).

Projection on the future in the person with SCD

In recent years, there has been significant progress in science concerning the treatment of chronic diseases, thus increasing the life expectancy of patients. One of the questions raised by these scientific and medical advances is the projection of the "sick" subject into the future. Few studies address this issue in adults with SCD. Buc-Caron & Galactéros (17) deal with it by emphasizing the professional career of people with this disease, leaving aside the way in which they envision their family and emotional future as people with this disease.

Pradere et al. (18) show that the vast majority of children with sickle cell disease find it difficult to plan for the future. These people always think of themselves in the present, a present marked by pain and by the anguish of death according to these authors who specify that the unpredictability of the crises, the device of the care and the frequency of the painful crises lead these affected children to show themselves pessimistic about their future.

In a socio-anthropological survey conducted in Niamey in Niger, Souley (19) specifies that most parents of children with sickle cell disease express themselves in religious terms: "I cannot envisage the future like that. Everything is in the hands of God..." Even if the parents consulted were divided on this point, we see that on the whole they cling to a certain hope rooted in their religious beliefs. They see "an uncertain future... When we see SS men living and working, it is a comfort... God alone knows if they will live, we don't know anything..." (19). These parents invest the future of their child with sickle cell disease with pessimism.

The pessimistic vision of the future of subjects with SCD can be grasped upstream from their school career. In Africa, Diop et al. (20) describe school careers punctuated by abandonment in children with sickle cell disease: 39.8% at the end of elementary school, 47.2% in middle school.

In France, the study by Buc-Caron & Galactéros (17) underlines the unpredictable nature of the disease, which makes it difficult for those affected to carry out personal and professional projects. They specify that the intensity of the professional activities arouses anxiety in the subject and plunges him into a situation of uncertainty in the face of the objective to be achieved, especially since his colleagues are not always informed of his condition. When they are, they do not necessarily understand the difficulties with which the patient is confronted. Added to this is the frequency of crises that cause absences that are sometimes unwelcome by some bosses. Moreover, these authors specify that even if subjects with sickle cell disease feel different from others, they mostly refuse the status of disabled worker, which could grant them working time and a position adapted to their situation.

The research of Njifon Nsangou & Scelles (10) on the children, broth-

ers and sisters of affected children encountered in Cameroon show that the latter project themselves on a future where professionals in the medico-social field, they will be able to overcome their helplessness, that of their family and their brother/sister reached by helping their parents nuance their care and nurture them. These research specify that they are also investing in a future where they will avoid having a child with sickle cell disease. What about adults with this disease?

Methodology

The research took place in a hospital environment.

Research Stance

The research was carried out in a Parisian hospital which receives patients mainly from Africa and the West Indies. To better understand and support patients, we went to meet them, at their bedside in the hospital to allow them to verbalize what they are going through regarding their disease and to express themselves as a person with SCD. This posture values them and makes them "experts" in their disease. During consultations, interviews with the psychologist, at the day hospital where bleeding and blood transfusions are carried out, during hospitalizations and in discussions with healthcare professionals, these patients were listened to in care's context. They were given the opportunity to express themselves and to say about their illness and the care as well as their quality of life. They were all receptive and agreed to participate in the research. This testifies to the interest they have given to this research.

Patients

For this research, the doctors, the psychologist and the nurses proposed to the researcher patients hospitalized in internal medicine or those who had come for their transfusion session at the day hospital by communicating to him their hospital room and/or their date of appointment. This testifies to the interest that this staff had for this research and to the research alliance between the caregivers and the researcher.

Twenty subjects from sub-Saharan Africa (12 women and 8 men), aged 19 to 71, including 10 born in France and 10 born in Africa, participated in this research. They have all been living in France for at least two months and express themselves well in French, the language spoken in the interviews.

Ethical devices

All participants gave their consent to participate in this research. They signed the consent form after an oral presentation of the object, objectives and research protocol which was made to them individually.

To ensure anonymity, fictitious first names were assigned to the patients. As provided for in the consent form for participation in the research, the interview tapes were destroyed after their transcription.

The data was collected via a semi-directive interview associated with a scale evaluating the quality of life in subjects. The semi-directive interview makes it possible to grasp the subjectivity of the subjects faced with a given situation. The interview is in fact a tool that questions the subject's relationship to the fact and not the fact itself (21).

The interviews were recorded on a Dictaphone with the consent of the patients. They took place in three different spaces: 4 in the psychologist's office, 6 in the day hospital during the transfusion sessions and 10 in the hospitalization rooms (9 in a single room and one in a twin room, in the absence of the second occupant of the room).

Before proposing an interview with the patient, the researcher first checked with the doctors, then with the patient about his state of health. When a patient was under the effect of morphine or pain, he was listened to but not included in the research.

The duration of the interviews varied between 1h and 1h30. At the day hospital and in the hospitalization rooms, interviews were often interrupted by caregivers who came to take the patient's parameters (tension, temperature, saturation, etc.). The researcher in fact ensured continuity of care despite the patient's participation in the research.

At the end of each interview, the patient was asked to complete the questionnaires, the WHOQOL-BREF. However, one of the patients returned their completed questionnaire two hours later and another, after a week, because of their level of French. Some, to complete this questionnaire, were helped by a nurse.

To evaluate the quality of life of the patients, we used the French version of the WHOQOL100 (Leplège et al., 2000) which evaluates the representations of the subject suffering from a chronic disease. The interview data were subject to a thematic analysis and the questionnaires according to an analysis inspired by Leplège et al. (22).

Results

The results are presented in the form of a cross-section of the experience of two groups of patients born in France (G1) and born in Africa (G2).

Representations of sickle cell disease

Patients give meaning to sickle cell disease by relying on knowledge from tradition and modernity.

“Western” biomedical representations

For subjects in both groups, sickle cell disease is a transmissible, genetic, hereditary and blood disease. They express themselves by borrowing a biomedical vocabulary. While the subjects of G1 do it easily and situate this disease on a medical level, those of G2 remain fixed on representations of the most prominent disease in their country of origin.

G1

In all subjects, the representations of sickle cell disease are based on its hereditary nature, which revolves around the transmission of the disease by the two healthy carrier parents. They recognize the responsibility of the parents on the transmission of the disease. Fatime specifies in this sense that “it is a hereditary disease which is transmitted by the parents, for example I am SS, if my husband is also SS we will necessarily have SS children. There, I am SS and my husband is AS, unfortunately my son is SS”. The disease is thus invested as a manifestation of bad luck in the affected subject and his family.

In addition to genes, the question of luck is put forward concerning the transmission of the disease. Therefore, one can be a carrier of the disease and, by a stroke of luck, not pass it on to the child. Marcus specifies that for a child to be affected “the parents must both be carriers of the genes. But then there is the question of luck unless both parents are SS. Otherwise if they are carriers there is the chance of also being a carrier or the bad luck of being sick. »

Adam recognizes the responsibility of his parents or his grandparents on the transmission of the disease. “It's hereditary but where it comes from, I don't know... My parents or a generation before because sometimes it skips a generation... but I know it's in the blood that it's trans-

mitted”. The origin of the disease is therefore strange to him, even if he knows that it is a hereditary disease.

G2

Alex invests in the search for the meaning and the origin of his illness. His crises make him remember those of his mother and his father who, like him, were suffering from SCD. “I started looking myself because I was curious, I was told it was a hereditary disease, certainly my parents had it... My father died in Dakar... he had anemia and by my curiosity I knew that people with sickle cell disease were often anemic. My father had sickle cell disease and my mother the same. That's why I have sickle cell disease!” He thus identifies with his parents who died of this disease and recognizes the hereditary origin of his disease.

Traditional representations

The subjects imagine SCD as a manifestation of the persecution of the patient by sorcerers. The subjects of G2 remain influenced by the representations that those around them have of sickle cell disease, unlike those of G1 who do not attach any interest to these representations.

G1

Patients say they know the traditional representations that members of their families, back home and/or here in France, have of sickle cell disease: incurable disease, coming from sorcerers, serious disease, disease that kills children at a young age. They say they do not share these representations of the disease. Jean emphasizes: “Like some in Africa who think it's witchcraft... It's true that to suddenly have a pain that you don't understand, you wonder what it's all about. ... maybe that's why other people often say it's witchcraft”. This verbatim therefore highlights the process of differentiation from the members of his family at stake in this father concerning the meaning given to the disease.

G2

All patients rely on representations conveyed in their country of origin to give meaning to their disease: shame, witchcraft, and curse.

Alex says that “in Africa we were starting to say, it's rheumatism ... at the time we said a lot of things, that it was witchcraft...» For Serge, “as they say in Africa in the village, they threw the disease on me, cast a spell...» The disease therefore has a cause external to the patient and is thus “thrown” on him by a malevolent being.

Moreover, it is a shameful disease which constitutes a taboo for the patient and his family.

Thérèse recognizes the unpredictable and chronic nature of her crises and highlights her difficulty in expressing her experience to her family and outside her family. Nina, for her part, says she suffers from the indifference of her family and extra-family relatives regarding the way in which she experiences her illness. She says: “The disease is not visible, so when you tell people that you are in pain, no one understands you, they ask themselves too many questions and I am ashamed of it”. The invisible nature of this disease is therefore a source of suffering for this woman.

Meaning given to sickle cell disease: between tradition and modernity

In both groups, all subjects draw on knowledge from both tradition and modernity to make sense of SCD. Even if they recognize the genetic nature of the disease, the strange nature of its attacks and the random nature of its transmission to the child lead them to see it as a manifestation of witchcraft. Bernadette specifies that sickle cell disease

is "strange... it wreaks a lot of havoc in Africa, many die of it at a young age and adults do not escape it". For Caroline, "it's a disease (silence), since my dad had it, so it's hereditary. But I always ask myself the question why me and not the others?". It is therefore a strange disease that leads to a lack of meaning in Caroline

Furthermore, patients see sickle cell disease as a manifestation of God's will and relate it to their destiny and that of the family. Odile declares "I tell myself somewhere it's destiny because I was born with it and that's what I call destiny... I tell myself since God gave it to me so I live with it". Thérèse expresses her powerlessness and resignation in the face of her illness. She says: "It is God who willed, it is he who gives and therefore nothing can be done against his will". These patients rely on the divine will to contain their suffering related to sickle cell disease and give it meaning. This "divine thought" is a resource in the face of the strangeness and suffering that sickle cell disease causes in these patients.

Choice of treatments by the subjects

The experience that subjects have of the disease in connection with the information they receive from those around them influences the choice they make of this or that treatment. In both groups, the subjects expressed a preference for the biomedical treatments deemed effective. They recognize the effectiveness of these pain treatments. Since sickle cell disease is a somatic disease, severe pain is relieved more quickly and better by biomedical treatment than by traditional treatment.

For some subjects, analgesic traditional treatments relieve physical and psychological pain. They act like "morphine" and are considered complementary to biomedical treatment. Indeed, these treatments (medicinal plants, scarification, amulet, etc.) remain a basis of security, even of maintenance in the family and the culture. They reinforce the containing function of the cultural envelope in these subjects. Thus, the meeting with the traditional healer can make it possible to reconstitute the unbalanced family group. Denise stresses in this regard: "I cannot cut myself off from my tradition". It also translates its investment in culture as a container and reassuring object in the face of illness. Culture therefore assumes a function of containing and transforming negative affects into a resource for this woman to adjust to her illness. By a rather remarkable slip, she speaks of "genius therapy" instead of "gene therapy". This genius can be understood as an "ancestral spirit" which heals and restores the vital dynamism of the patient and the group. The entourage and the family group play a key role in the implementation of the traditional therapy. It is a kind of mothering, of support that is put in place to support and maintain the link with the subject regardless of the distance (23).

Unlike subjects born in France, those born in Africa show more resistance to exchange transfusions. This resistance can be understood by the fact that they are afraid of being contaminated by blood. Infections such as malaria and viral hepatitis are quite common, but the most feared remains HIV-AIDS (24); in Africa, many patients have been victims, contaminated through blood transfusion (Bonnet, 2001). Moreover, the blood represents life and the other. The subjects are often afraid of being possessed by another, an unknown by his blood, a blood perhaps dirty, impure, and thus they fear losing their identity. There are therefore many interpretations around transfusion exchanges which are very often a source of concern for patients and their

families. This leads to non-compliance with this treatment by patients and their families.

The subjects who were born in France accept and desire the transfusion which, according to them, is necessary for their well-being. The positive experience they have had since childhood in France with transfusion reinforces their fairly good compliance with this treatment. Moreover, they grew up in a context far from the African symbolism around blood and they are sensitive to the guarantees of safety offered by conventional Western medicine concerning transfusion.

Projection of patients on the future

The future is considered in connection with the choice of profession, parenthood and/or country of residence.

Optimism/pessimism about the future

The future of patients is thought out by them, depending on the case, with optimism or pessimism.

G1

Several trends emerged regarding sentiment towards the future. Only 3 out of 10 patients remain optimistic about their future. Odile specifies: "I think we can live well with it, the proof, I have my son, in 4 months, and I will graduate. I am confident, I have more confidence in the future", Marcus says he is "Serene, but with a point of uncertainty... in relation to the illness, yes, everything is fine and all of a sudden big crises... I am calm because I want to get out of it but the disease is there when I lower my vigilance..." For him, illness is an obstacle to the realization of oneself and one's projects. For Martin, on the contrary, "SCD does not prevent me from doing what I want to do".

Two patients want to overcome the handicap of sickle cell disease in carrying out their projects. Assa is aware and refuses to resign herself to the handicap that her illness constitutes for her future. She specifies that "the SCD causes us a lot of problems... the disease can never slow down my projects. It can slow me down at the time of the crisis, but that's not why I'm going to close the doors. For Mariam "I want to get better, have a more or less normal life, what... to travel to other countries".

Other patients find it difficult to detach themselves from the present to invest in the future. They remain fixed in a present marked by painful crises and suffer from what those around them think and say about their illness and themselves. Olivier often heard members of his extended family tell his parents that he would not live to be 16. This leads him to live daily with a fear of death that hinders his ability to serenely invest in the future. "I can't plan for the future... Because as a patient, it's hard to plan. I'm not going to grow old, I'd rather die." The future is therefore thought of in terms of imminent and inevitable death for this young man.

The chronic and lethal nature of Marie's crises leads her to avoid thinking about her future. This future remains "a bit vague... Today, it's a bit messy, I don't know..." It is therefore a future that presents itself with strangeness in this woman.

G2

All patients are pessimistic about their future. They identify with their family members and those around them who died of sickle cell crises. This arouses in them a fear of death in connection with their illness. Denise declares: "It's hard... because sometimes I think that maybe I'm going to die, I tell myself a lot of things: I want to see my children grow

up... What really struck me was that my aunt, my mother's little sister died at 42. She is thus afraid of dying like her aunt.

François remains influenced by the relatively short life expectancy of people with sickle cell disease in his home country. He points out that "Considering my situation today, I hope I will be dead... sickle cell patients do not have a long enough life expectancy, maybe in the future science will do something. But what I've heard around me, what I've read, and life expectancy is 40 to 50 years, but sometimes I don't think about it... it slows me down." He therefore avoids thinking about the future, a future that he finds difficult to dissociate from his own imminent death.

Contribution of the Beck Despair Scale

All subjects completed the questionnaire. Twelve subjects out of 20 asked for an explanation of items 8 ("I expect to have more good things in life than the average person") and 9 ("I have no respite and there is no reason to believe that I will have any in the future"). This request can be linked to their desire to fully understand these questions in order to better express their experience and communicate it to the researcher. It can also provide information on the research alliance between the participants and the researcher.

In this questionnaire, a higher score predicts a negative feeling in the future. The results obtained at the BHS show that patients born in Africa have an average score of 6.9 while those born in France have a score of 4. Thus, those born in Africa face the future with more despair than those born in France. These results are consistent with those from the interviews.

Question of parenthood in patients

The desire for parenthood is present in all patients.

G1

In this group, 3 patients are already parents and no longer wish to have other children because of the difficulties encountered with their children. Odile says: "I'm afraid of having another child, a pregnancy... I had barely given birth when my baby was taken away from me... when the crisis occurred, but in itself, it's an episode that traumatized me and still traumatizes me... for the moment, I don't want to have a child". The other patients (7) wish to be parents. The number of children desired by these patients varies between 1 and 3. They say they are not concerned about the status of their partner.

Assa is the only patient who says she demanded premarital exams from her husband. She specifies that: "My companion is AA, I wanted him to do the blood test, and he tells me what we do if it is positive? We break up! I'm SS, I don't want to make people unhappy. I told him a sentence that touched him: even if we love each other, we must not make our descendants suffer, so he took the test, everything went well..." So she did it to protect her family and children from the suffering she is going through with sickle cell disease.

G2

The desire for children and/or an unaffected spouse is present in 8 subjects out of 10. Two say they want to be very careful about the choice of partner. Adele wants to marry a spouse who does not have sickle cell disease and have children like her mother.

Six of the subjects live as a couple with or without children, and 2 of them, older (Christine and Alex) are grandparents. Two women say they have difficulty conceiving a child. Nina says: "We tried several

times, it didn't work because of the disease... if I really can't do it, and I'm going to adopt a child. Children are important." For Caroline, "the project of having children fell through because I had an early menopause... I had miscarriages all because of my illness". The disease is therefore experienced by this woman as an obstacle to her process of parenthood.

Two of the youngest patients (24 years old) are pessimistic and sometimes ambivalent about parenthood. François declares: "I don't have the idea of marriage, I don't think I'm cut out to get married... Because I don't know how to raise my children, or if they had my disease, what would I do? I would like to have 2 or 3 children, to be in a couple, but I can't imagine myself at this stage.". According to Thérèse, "If you are in a relationship, the man will want you to get pregnant for him. No, I'm not ready... it's not possible, even in five years." Two patients remain optimistic, but with uncertainty about whether or not they will be accepted by a partner. Serge suffers from not being able to talk about his illness directly/openly to a woman. He is actually afraid of being rejected by her if she comes to know that he has sickle cell disease. Indeed, he is looking for a French spouse. "I am looking to marry a French woman". For him, therefore, a French woman will be more understanding and more supportive than an African woman in his confrontation with his illness.

Projection in the professional world

The subjects project themselves into the future by considering exercising various professions, often related to their disease.

G1

The questioning relating to the professional future is present in all the subjects (being a doctor or an electrical business manager, being a civil servant, etc.). Those who have stopped their studies wish to resume them in order to have a job. Marcus wants to get his bachelor's degree to become a computer network administrator. Marie would like to help people with sickle cell disease by becoming a doctor. She sees herself "in any case working at the hospital". She therefore wishes to put her experience to the benefit of people in suffering and/or hospitalized.

Patients who already have professional activities say they have chosen them taking into account their illness and the requirements of their parents. They acknowledge having been influenced in their choice by their parents who overprotect them by guiding them towards professions that do not require too much physical effort.

G2

Eight of the 10 subjects in this group say they have been influenced in their professional choices by parents, by illness and for one of them by a doctor. All of them say they first wanted to work in the health field as doctors, nurses, caregivers. For Thérèse, "Medicine is my passion... I spoke to my doctor about it, he told me to be honest with you, and you won't be able to do medicine... it's not easy studies... I don't I don't have the health to do all that...my parents...it's better to follow your doctor's advice! It's true, given the large number of seizures I had when I was at my parents' house". This verbatim provides information on the suffering caused in this woman by her difficulties in pursuing advanced studies in order to be able to realize her dream of becoming a doctor. This handicap, due to his illness, is experienced with helplessness. She tries to contain this suffering by rationalizing and trying to

follow the doctor's advice, despite herself.

Three patients want to get involved in humanitarian action and work on their own. Adèle dreams of being a humanitarian entrepreneur and says: "to be my own boss, to do what I want to do. I don't want a boss who's going to tell me do this, do that." She expresses, beyond her desire to help others, her desire for autonomy in relation to others and to her illness which impose on her a lifestyle to which she has difficulty adjusting.

Discussion

The research highlights the coexistence of 2 systems of representation of sickle cell disease, traditional representations and biomedical representations both in patients born in Africa and in those born in France. They therefore rely on knowledge from tradition and modernity to give meaning to sickle cell disease. This result is consistent with those of the literature (14) and can be understood by the fact that in African families or families originating from Africa, the tradition has neither passed nor been exceeded (5).

Subjects born in Africa show a tendency more marked by traditional representations of SCD, without however turning away from western representations of this disease. Among those born in France, the trend is reversed. This can be explained by the intricacy of the social and the psychological concerning this disease. The subjective experience of sickle cell disease by a subject and the beliefs shared by those around him therefore influence the representation of the disease. Despite the level of integration of the subjects in France, they have not completely broken with the reality of their country of origin (culture, mortality, dysfunction of the healthcare system, etc.) (7). They are still influenced by what is said and thought about sickle cell disease in their countries of origin. They have in fact internalized traditional representations of sickle cell disease without rejecting modern/Western representations. The theme of religion emerged in the discourse of the subjects, an object which seems essential in the understanding and experience of the disease in the subjects of the two groups. They are strongly marked by the belief in the divine and predestination. Everything that happens in life is the manifestation of the will of God: it must be accepted. Religion then becomes a coping strategy that allows subjects to give meaning to their illness and to accept it (8). They thus contain their psychic suffering and despite everything hope to live as long as possible because the divine will help them to find healing. We are witnessing, in these patients, a kind of psychotherapy of God as thought by Cyrulnik (25).

All subjects born in Africa manifest a "therapeutic wandering". They have recourse to the traditional healer, the doctor and the divinity at the same time to relieve the attacks. This wandering constitutes, for them, a mechanism of identity reconstruction (13). Their therapeutic path is related to their representations of SCD, influenced by the singular experience and the information received concerning this disease. Sickle cell disease remains in fact a subjective experience imbued with the cultural and environmental reality in which the patient and his family live (26).

The question of death is significant in this research. SCD is considered an incurable disease by patients born in Africa and constitutes a fatality, "a path of no return". This nourishes, in these patients, on a daily basis, an omnipresence of death. This tragic, negative experi-

ence of the disease does not promote "luck" or the possibility of hoping to "taste" life. This prevents the patient from realizing himself and thinking about his future with serenity. Patients born in Africa have witnessed the death of people with sickle cell disease or heard about it from relatives who considered them in danger of death, who thought of them as subjects whose death was imminent. This experience arouses in them an anguish of imminent death despite the quality of the care they receive in France. Chronic pain obstructs their projection into the future. The score obtained by these patients on the BHS scale shows their tendency to demotivation, to pessimism concerning the investment of their future.

Patients born in France think of their disease as chronic and not fatal. If death anxiety is present in these patients, they do not feel the imminence of their own death acutely, unlike patients born in Africa. Therefore, they project themselves on the future with less pessimism than patients born in Africa.

Moreover, all the subjects expressed a desire for a child despite their knowledge of the mode of transmission of the disease. This can be understood by the fact that the human being is marked by the desire to ensure his descendants. The subject, whatever his state, thinks about his survival, that of his family and his lineage: for the African subject, the child is a wealth and a source of social valorization for the parents and their families. Procreation is, therefore, an impetus of life that inscribes the subject in socio-cultural "normality". All patients have the representation of the disease "who is there and who is not there (absence vs presence)". Giving birth to children is a source of social valorization and psychological well-being for patients in a socio-cultural context where the status of men and women depends on their ability to be fathers or mothers (16).

Conclusion

This research conducted with adult patients followed in a Parisian hospital presents results similar to those of Scelles (27) on the meaning of the choice of medico-social professions for a woman, sister of a child with intellectual disability. She also presents results similar to those of Njifon Nsangou & Scelles (10) on the coexistence, in siblings of children with sickle cell disease, of two explanatory systems for sickle cell disease, modernity and tradition. Concerning the projection on the future, it presents results similar to those of Buc-Caron & Galactéros (17) concerning the difficulties encountered by adults with sickle cell disease in their professional environment. This research made it possible to dive into the world of representations that subjects with sickle cell disease and living in France have about this disease, whether they were born in France or in Africa. She highlighted the advantage that patients born and cared for in France have over those born in Africa and cared for or not there in terms of their quality of life, with considerable implications for the quality of their projection on the future. The research highlights the issue of the cultural, social and medical environment on the process of subjectivation of sickle cell disease by affected adults. It has made it possible to highlight the "individual experience" of the disease and the "original individual representations" constituted in "profane theory" (28). The passage from one culture to another is disturbing, it is therefore necessary that a cultural mediator and a psychologist trained in intercultural matters intervene in the care system; the first to facilitate the transition from one culture to

another with respect for each person and their values, the second to ease the tensions inherent in this transition.

Sickle cell disease is a disabling disease: painful crises, acute and chronic complications, hospitalizations throughout the patient's life can be a source of anxiety, discouragement, depression in the patient and his relatives. The gaze of others, the experience of difference, the difficulty of finding one's place in society are all challenges to be met by the patient and caregivers.

These psychic and cultural problems cannot be solved by simple therapeutic education or counseling of patients and their relatives. Transcultural consultation, in this context, can bring to light the contradictions between the representations and symbols of the patient's culture and the rational information given by conventional Western medicine, contradictions at the origin of certain disorders in the patient. By explaining the data of conventional Western medicine and recognizing the cultural representations of the patient, we can achieve a certain clarification, a meaning of the data which in itself has a therapeutic value for the patient. This approach can be an efficient recourse for patients and their families with strong cultural roots. Articulating the different representations could help patients to talk, to think about their life, to understand themselves and to understand what is happening to them in order to live their situation and project themselves into the future with more security.

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