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Assessment Specificities of Cameroonian and Psychological Experience of Brothers and Sisters of Children Affected by Sickle Cell Disease

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Abstract

Sickle-cell disease is the most widespread genetic disease in the world, and it confronts the sick child and his or her family with painful, unpredictable and chronic crises, multiple hospitalisations that are costly for the family, and the thought of imminent death. The disease is a taboo and a trauma in Cameroonian families, where only children are rare. This article examines the experiences of siblings of children with sickle cell disease in Cameroonian families where this disease is a taboo and where the meaning given to the disease is based on both modern and traditional references. Its aim is to improve the psychological care of children with the disease and their families by taking into account the psychological relationship that siblings and families of children affected with the disease have with this disease, the care of the sick child and the sick child himself.

Keywords: Siblings; Family; Sibling; Sickle cell disease; Cameroon

Introduction

Sickle cell disease is the most common genetic disease in the world. It is a disease which confronts families with the child's repeated, intense and unpredictable crises, and with his or her death, thought to be imminent and inevitable, which is a taboo in Cameroonian families where it is difficult to find families with an only child. The brothers and sisters of children with sickle cell disease witness their crises on a daily basis and the behaviour of their family members in the face of these crises. They see their brother/sister transform during crises in a family context where sickle cell disease is taboo. This research examines the experiences of the brothers and sisters of children with sickle cell disease with regard to the disease, the child with the disease and the child's care.

General information on sickle cell disease

Sickle cell disease (SCD) is a genetic, chronic, lethal and taboo disease in Africa. It is the most common genetic disease in the world, with around 500 million people carrying the sickle cell trait and 50 million affected worldwide [1]. Initially found in malaria-endemic areas such as sub-Saharan Africa, migratory movements of populations have gradually modified its distribution around the world. Since then, the disease has been present in virtually all countries with populations originating from Africa and the Mediterranean region [2]. Unlike in most Western countries, such as France, where it is a rare disease [3], SCD is highly prevalent in almost all African countries. Africa is the continent most affected by SCD, with a prevalence rate of 2% in the general population and an estimated mortality rate of over 70% in children under 5 [4]. With a prevalence rate of 8.34% in the general population, Cameroon is one of the countries most affected by this disease [5]. In Africa, SCD is expensive to treat, and the unavailability of bone marrow transplants and gene therapy, the only effective treatment for the disease, means that many children under the age of 5 die [6]. Discovered in the United States of America in 1910, SCD is an autosomal recessive disease transmitted to children by both parents. It is caused by the presence of abnormal haemoglobin in the blood, which results in a shortage of oxygen to the body's various organs via the red blood cells, which have a reduced lifespan. The rapid destruction of red blood cells leads to anaemia and chronic, unpredictable

pain with which the patient identifies himself [7], resulting in multiple hospitalisations and costly treatment for the parents of affected children [8], who often have a traditional representation of the disease.

The specificities of families adjustment with SCD

In Cameroonian families, sickle cell disease is seen in terms of persecution, guilt and taboo.

Sickle cell disease, a figure of persecution for familie

In the cultural context of sub-Saharan Africa, illness is seen as an "evil" capable of attacking all members of the family, even after the death of the patient [9]. Sick children live in a society structured by traditional prohibitions, rites and attitudes, of which women are the guardians [10]. A child's illness or disability is part of this cultural structure, which gives it meaning and produces effects. SCD is seen as a manifestation of the possession of the sick child and his family by a sorcerer or evil spirit [11]. Depending on the case, it may also be experienced as a request from the ancestors to the patient's family to make amends for a transgression of an ancestral norm [11]. Generally, the mother is held responsible for this transgression; the over-protection of the affected child, by her and by the members of the family, constitutes a defence enabling them to relieve themselves of guilt and to inscribe the sick child in the history of the family [1].

SCD: a family taboo

Talking about SCD is taboo in African families or families of African origin [2]. As a result, children suffering from this disease often have difficulty talking about their illness and verbalising their experiences [12]. Parents hide the name of the disease from some or all of their children, and children hide their knowledge of the disease from their parents [13]. For the children, this taboo is a means of protecting their parents, and for the parents it is a desire not to add to their children's suffering. Parents also use this taboo as a means of protecting their sick children from social and family stigmatisation [13]. Parents are pessimistic about their child's future and want to continue to invest that future with hope. They may therefore think that not talking about the disease protect the children. This way, they develop a family fantasy to protect their child from this disastrous future and from the death of this child, thought to be imminent and unpredictable [14].

Specific features of the family and siblings in Cameroon

The African family is a highly complex reality that transcends biological considerations. It is based more on the social rules of alliance and filiation than on blood ties, biology and parenthood. The family is built around a set of values and norms. The Cameroonian family is essentially thought of as an ethical reference based on the subject's feeling of belonging to a group. As such, it is trans-spatial and transcultural. A Cameroonian may have a village family, a city family, a university family or a neighbourhood family [11]. From the "family spirit" obviously flows what is known as the "sibling spirit". As a result, a friend, a classmate or the child of a family friend is often considered a brother in the African context. This makes African siblings, in general, and Cameroonian siblings, in particular, complex. This complexity can be understood by the fact that in Cameroon, those who educate, protect or in any way promote the subject's personal fulfilment are often considered to be family members. However, the Cameroonian family has not withstood the social changes brought about by its encounter with the Western world (colonisation, globalisation). African families have preserved certain cultural values such as polygamy and the maintenance of the life of the family group through the birth of several children; "life is man" [15]. They have also opened up to modernity by gradually applying family planning. In Cameroonian, tradition is neither passed nor outdated [11]. Individual behaviour is still marked by identifiable traditional elements. These families have neither resisted nor surrendered to modernity. They are at once modern and traditional, neither modern nor traditional. They are therefore in the middle ground between modernity and tradition. The parents of the affected child, supported by the members of their respective extended families, seek ways to alleviate the child's suffering. With the advent of globalisation, they turn to a number of therapies. The therapeutic itinerary is, in this sense, a sort of mosaic between traditional therapies and imported therapies such as Western therapies and messianic, religious therapies, so to speak [16]. The traditional therapist, the doctor and the imam, pastor or priest are consulted, usually at the same time. The system of "care in the round" [17] perfectly sums up the therapeutic path taken by most sub-Saharan African families. With each therapist, they seek to heal the child's specific suffering. In fact, the quest for meaning in the child's illness and the quest for ancestral protection and reconciliation with the ancestors lead them to the traditional therapists. With the priest, the pastor or the Imam, they seek divine therapy to ensure the protection of the Supreme Being. They generally do this because they want the child to be cured under God's mercy. With the doctor, they seek the child's somatic healing. These families are on a permanent quest to rebuild their identity [18]. The identity of Africans remains highly complex. They find it hard to define themselves; they are neither westernised nor African. As a result of the effects of globalisation, Cameroon is constantly seeking its own identity [11]. There are several types of family in Cameroon: nuclear families, extended families, polygamous families and monogamous families [19]. So Cameroonian families are both traditional and modern. This reflects the diversity and complexity of Cameroonian families. These different types of family give rise to different types of siblings (blended, large, nuclear).

Talking about sickle cell disease in the siblings of a child with the disease

Children learn from their parents a way of being and being a sibling; they also understand what it is appropriate to do, say and think as a family, and again, by means of explicit and more often than not implicit prohibitions, they learn to keep quiet about some things and not to think about others [20]. The brothers and sisters of children with sickle cell disease keep their suffering about their sibling's disability a secret from their parents, the sick child and other siblings. They do so, supported by the family taboo maintained by the parents around the disability, so as not to exacerbate the suffering of those to whom this suffering is not told [2]. For these children, this taboo is a sign of recognition of the suffering of the members of the family group and a specific way of dealing with this suffering, which is then marked by emotional isolation. Every sibling, whether or not they include a disabled child, is the fruit of a weaving process in which the parents have provided both the weave and the technique. The parents' history and culture lead them to pass on to their children what it means to be "a brother", "a sister", "an elder", "a younger". Based on this "know-how" and "know-how to be", each child, interacting with the dynamics of his or her peer group, first applies the "basic technique" and then gradually develops it further [20].

The feelings experienced by the brothers and sisters of the sick child

We met sisters and brothers (aged of 8-19 years old) of children with sickle cell disease in Cameroun with the aim to describe their complex feelings about their sick child and his or her illness. The analysis of interviews gave following results.

The painful feeling of strangeness and isolation

Because of the family taboo on SCD, the sick child and his or her illness are a stranger to brothers and sisters. They find it hard to think about him and to imagine him. Brothers and sisters do not understand what the patient is going through. The strangeness of the patient hinders the siblings' process of identification and differentiation with the patient, and makes them feel powerless and angry at the patient for not telling them what he or she is going through with the disease. The suffering of the brothers and sisters of children with SCD lies in the inability of the latter to tell the former what they are feeling and experiencing as a result of their illness. For the brothers and sisters, this suffering reflects their desire to have power over their brother's illness, to give it meaning through a precise representation of his attacks. It also reflects their impaired projective identification with the patient. The patient remains a strange person who causes them suffering and a feeling of powerlessness. Obviously, not understanding what the sick brother is going through with his illness, not understanding what is happening in his body, not being able to imagine his attacks, leads the brothers and sisters to ask questions about themselves, about their own identity, when we know that children build their identity through processes of identification and differentiation from others. SCD, through its crises, alters the process of identification and differentiation with the patient in the siblings. This causes them psychological suffering. The brothers and sisters of children with sickle cell disease, parentified by their parents in monitoring the child and preventing crises, experience a feeling of emotional isolation. This feeling is due to the fact that they are not given the opportunity to tell the patient, the brothers and sisters who are not sick, or the parents, what they are experiencing in terms of this way of being and being a sibling with the sick child.

The complex feeling of guilt

The children's experience of guilt is complex. Doriane feels responsible for Marc's illness and seizures, on the one hand because her mother had warned her about the risks and she had failed to keep a close eye on her siblings, and on the other because she feels guilty for resenting this vulnerable child. Joseph sees himself as both a victim and a perpetrator of Mark's illness and behaviour. Partly as a result of this guilt, Doriane and Joseph are angry with Marc, who occupies a central place in the family, who causes them torment and who puts them in the position of not knowing/being able to be a good sister and a good daughter/son. The siblings, as a result of their aggressive behaviour towards the child with the disorder, often believe themselves to be primarily responsible for the child's outbursts. Doriane feels responsible for her Brother Marc's seizures, which she associates with her own aggressive behaviour towards him. "When I hit him, my anger diminishes. But when his illness starts up again, it's as if it's because I hit him that his illness starts up again", she says. She knows that she has transgressed the family norm of protecting Marc in a cultural context where it is the daughter's duty to look after her brothers and sisters. While this aggression temporarily soothes her, it gives rise to painful guilt. If they feel guilty, the brothers and sisters also feel that the adults are "making" them guilty for things for which they alone are responsible. They also know that the adult, unhappy and powerless, can unleash anger and aggression on children who are not ill, which the child has to accept without being able to fight back. Doriane’s visits to her brother's hospital led her to realise that the disease is transmitted genetically to the child by both parents. She recognises that the mother is responsible for the disease. As a result, she refuses to be designated by the mother as responsible for her brother's illness and believes that the mother should assume her responsibilities in caring for the patient. In this way, she refused to be parentified by her mother. Girls were more

involved than boys in caring for their sick child. This difference is linked to the fact that the education system for girls in Cameroonian families aims to make them women capable of taking good care of their children's health and education. While brothers and sisters feel abandoned by their families, sisters are less likely to express their feelings of loneliness. This leads to differences in the children's experiences of the disease and the child with it. As a result, the sisters feel more responsible for the crises, unlike the brothers who feel more victimised by them.

The worrying question of death

The question of death is very present among the children, who raise it without too much resistance. They talk about the fact that their sick brother might die, with the idea that this death might not make the illness go away. Indeed, if their brother dies, the disease could affect them. The ideas that unaffected children have about death often stem from their curiosity about what adults have to say about the disease and/or the affected brother or sister. Ornelle, the older sister of a child with the disease, whom she often assists during hospital stays, explains: "The doctors have said that he's going to stay with his disease until the end. He's going to die with his disease. The other day, when my uncle came to the house, he also said that the disease believes that if it kills Marc, it won't also die with him". This "transfer" of the illness can occur when the affected child dies or when he or she recovers. But as recovery is not envisaged, only death leads to another child embodying the disease. She says this even though she knows that the disease is genetic and not "contagious" in the strict sense of the term. What she knows about the "medical reality" of the disease coexists with the meaning and functions that her culture attributes to this "disease". Children are afraid not only for the patient but also, probably, for themselves, in a complex process of identification. Children have difficulty coping with the fear they experience on a daily basis because of the iterative, unpredictable and severe nature of their brother's or sister's attacks.Ornella, talking about the fear she experiences regarding her brother with the disease, says: "It's always with me, even when he's well. I know that everything can change in a single second". So she never feels secured, always on the alert, even in hospital. She includes the patient's death in this "everything". Unaffected children express a complex desire for the death of their affected child. This desire is based both on reality (expenditure on care, fear generated by strange seizures) and fantasies (he won't survive). These children therefore oscillate between the desire and the fear of death for the sick child who, despite his illness, remains 'other', the same; to desire his death is, ultimately, to evoke one's own death in a process of self/other confusion, generally at work in siblings [21]. Moreover, the physiological/body transformations of the sick child during his seizures arouse or reactivate in his brothers and sisters the fear/desire of death but also a feeling of strangeness in the face of this 'same as oneself' who is being transformed.

The question of care and the relationship with care

Sick children are treated using both traditional African medicine and conventional Western medicine. Although complementary, these medical practices are experienced differently by the siblings

Conventional Western medicine

The hospital is seen as having a curative function for the seizures of the affected child, given as a matter of urgency. Generally, the family only seeks hospital care after self-medication has failed and preventive measures have been implemented to avoid or relieve the child's seizures. Obviously, this behaviour on the part of the family is an adaptation to the costly nature of care in a context where care is not reimbursed and health insurance is not accessible [22] to most families. This poses the problem of preventing seizures by children with SCD attending day hospitals in sub-Saharan Africa in general, and in Cameroon in particular. Seizures in children with sickle cell disease are treated in hospital by transfusion, which relieves them for a time without eliminating them. Only the affected child receives medical care. The children recognise the effectiveness of this treatment in temporarily relieving their child's attacks. This arouses and/or reinforces their fear of death. Paulette's illness is thought of as a "blood disease" by Jules, her younger brother, who believes that the "sick" blood should be drained from Paulette's body. "For it to end, you have to drain all the blood from your body. That's how you get rid of the disease". In his opinion, hospital care professionals do not do this, because they put new blood into the sick child's body and do nothing about the "bad" blood in her body. Hospital care professionals are designated by Jules as the "responsible party" for the chronicity of the crises and disqualified from the process of definitive relief of these crises. This recognition of the ineffectiveness of hospital care arouses his anger at these professionals, whom he sees as ineffective in preventing his sister's worsening condition. It's possible that through this anger, Jules is projecting onto these professionals his feelings of powerlessness regarding his sister's crises and the concomitant feelings of guilt. It can also be understood as a demand for recognition and acknowledgement of his experiences by medical professionals. Siblings also feel rejected and disregarded by medical professionals, who focus their care on the affected child. This situation arouses in them a feeling of anger towards the carers. This feeling is a sign of their desire not only to be recognised as brothers and sisters of the child with the disease, but also to be cared for and supported as such by medical staff.

Traditional African Medicine

Unlike conventional Western hospital carers, traditional practitioners care for both the child with SCD and his or her siblings. The sick child, his brothers and sisters, his parents and extended family are cared for by the traditional healer because the illness is seen as the illness of the patient and his family. The traditional healer treats the patient's body, which is thought to be possessed by 'evil', an evil that manifests itself in the patient as chronic disease and attacks of pain. These symptoms tell the traditional healer about the extent of possession and persecution of the sick child's body by the sorcerers. In response to this possession/persecution, the traditional healer administers bodily care to the patient and members of his family. For the siblings of the sick child, there is a before and an after to taking the traditional treatment. A "non-secure" before, where the threat of contamination by the disease looms large, and a more "secure" after, where this threat is contained by taking the traditional treatment. The children experience this treatment as protection against contamination by the disease. Taking this treatment is associated with a reduction in fear about the disease and being infected by it. However, the fear of death in the sick child did not disappear in his brothers and sisters after taking the traditional treatment; it was reduced. This treatment has led them to experience their bodies as less vulnerable and more resistant to the disease. As a result, they feel safer from the disease and from the witch doctors. From then on, this treatment helped them to contain their feelings of persecution by the witch doctors and their fear of being infected by the disease, but did not eliminate this fear. Talking about his sister's traditional care, Jules says that "wounds were made on her body". Parts of her body were scarified. Scarification of the painful parts of the patient's body allows the therapist to act directly on the evil, to limit the destruction of the patient's body by the sorcerers. By scarifying the painful parts of the patient's body, the therapist administers a powder with magical powers that is effective against sorcerers. From then on, the aim of treating the patient was curative, with the aim of "freeing" the patient from possession and persecution by the witches. So it's not just the blood that's treated here, but also the body. Paulette was accompanied to the traditional practitioner by her mother and her paternal uncle. Her brothers and sisters, who were not present at the consultation, received the treatment given by the traditional therapist through their uncle. Jules mentioned the fact that his maternal uncle had scarified him and his other siblings, who did not have sickle cell disease, with the powder given to him by the traditional practitioner. As the father had died, it was the paternal uncle who went with the mother and the sick child to the tradi-pratician's consultation. This shows the involvement of the extended family in traditional care and supports the mother in this approach. The uncle is also an intermediary between the traditional practitioner and the family. He is responsible for He is responsible for supervising the application of traditional treatments. Like the sick sister, Jules was scarified by his paternal uncle, at the request of the traditional healer. The treatment was administered orally and by scarification of the back and sell. "Even my mother and older sister ate the same medicine. We ate and then the rest was put in our blood". Paulette's brother, sister and mother, who were considered to be at risk of becoming ill and being persecuted by witch doctors, were also treated for sickle cell disease by the traditional healer. The aim of this treatment is to protect them against the disease. For Jules, the patient's blood is dead blood, blood 'possessed' by sorcerers. In this sense, it is "bad blood" that attacks the "good blood" transfused to the patient during hospitalisation following anaemic crises. According to him, this explains the chronic anaemia and recurring bouts of pain suffered by his sick sister. The traditional treatment is thus thought to be effective against sorcerers. It makes the patient's blood and that of non-patients unassailable by the sorcerers. Taking it reduces the feeling of persecution by the witches, even if the sick sister dies. "They won't look for me anymore", he says. The traditional treatment is thus seen as protection against attacks by witches, against illness and, of course, against the death of the sick sister and against his own death. Taking the traditional treatment leads Jules to say of the sorcerers that "It's all over for them, everyone has eaten the remedy, everyone is armoured", and to specify that "When the sorcerers enter the family, they don't come out any more". This ambivalence in relation to the traditional treatment (protection and vulnerability) in the face of witches suggests that the feeling of security against witches that Jules talks about remains apparent, that he is constantly experiencing a feeling of insecurity that is never really eliminated.

Conclusion

This research shows the need for professionals to remain extremely sensitive to the way in which the experience of the disease is embedded in the family and sibling psyche of children confronted with sickle cell disease in their brother/sister in Cameroon. Taking into account the clinical and cultural data presented here can help to make the family and siblings a resource for the child with SCD, his/her parents and siblings, as well as for carers confronted with this lethal disease. Discussion spaces between the sick child, his or her brothers and sisters, parents and healthcare professionals within institutions can help to achieve this objective and reduce the potentially harmful effect of the family taboo on sickle cell disease on the sick child, his or her brothers and sisters and even his or her parents. The denial pact can have a protective effect, temporarily, for the sick child, his parents and siblings, if it does not last too long. However, this space for discussion should be built by all the participants, who should (beforehand or gradually) understand the benefits for each of them.

References

1. Nsangou HN. Sickle cell anemia and the mother-child dyad in Cameroon. Cah Childcare worker. 2022; 59: 31-33.
2. Nsangou HN, Scelles R. Sickle Cell Anemia and Family taboo: The experience of a Sister of a Sick Child in Cameroon. Am J Pediatr. 2020; 6: 190-198.
3. Nsangou HN, Scelles R. Sickle cell anemia and projection towards the future: a comprehensive approach to the experience of a sister of an affected child in Cameroon. Neuropsychiatr Childhood Adolescents. 2020; 68: 410-420.
4. O World Health, Training manual on strategic planning for cervical cancer prevention and control in Africa. 2017.
5. Weatherall DJ, Clegg JB. Inherited haemoglobin disorders: an increasing global health problem. Bull World Health Organ. 2001; 79: 704-712.
6. Drain E, Pradere J, Taieb O, Dutray B, Champion M, Bonnet D, et al. Healing process of chronic disease: sickle cell disease treated with allogeneic hematopoietic stem cell transplantation: main results in adolescents. SFPEADA National Days. French Society of Child and Adolescent Psychiatry and Associated Disciplines, publisher. Neuropsychiatr Childhood Adolescents. 2008; 56: 305-310.
7. Josset-Raffet E, Yi MK, Benkerrou M. The bodily and psychological trajectory of pain in children with sickle cell disease. Neuropsychiatr Childhood Adolescents. 2016; 64: 131-138.
8. Nsangou HN, Scelles R. The Psychological Impact of Sickle Cell Disease on the Sick Child's Family. Acta Sci Paediatr. 2020; 3: 39-47.
9. Njifon Nsangou H, Scelles R. Sickle cell anemia and siblings: a look at the experiences of a sister and a brother of a sick child. J Pediatric Childcare. 2019; 32: 75-84.
10. Keita T. Traditional phenomenology of childhood in Africa. In: Itoua F, Tettekpoe DA, Traore A, Keita T, M'Baye M, Kotto E, et al. publishers. Family, child and development in Africa. Paris: Uneso. 1988. 99-139.
11. Tsala Tsala JP. African families in therapy: Cameroonian Family Clinic. Paris: Editions L'Harmattan; 2009; 268.
12. Nsangou HN, Falck J, Scelles R. Family culture of sickle cell disease and body image in affected children. Ann Med-Psychol Rev Psychiatr. 2020; 178: 827-836.
13. Bonnet D. Breaking of alliance versus breaking of filiation: the case of sickle cell anemia. In: Dozon JP, Fassin D, editeurs. Criticism of public health: an anthropological approach. Paris: Balland. 2001; 257-280.
14. Souley A. Discourse around sickle cell anemia in the Hausa community in Niger. In: Laine A, Bonnet D, Keclard L, Romana M, editeurs. Sickle Cell Disease: Perspectives on an orphan disease. Paris: Karthala; 2004; 141-69.
15. Yao KM. Family and parenthood in Africa at a time of societal change. Paris: L'Harmattan. 2014. 270.
16. Nsangou HN, Scelles R. Sickle Cell Anemia, Representations and Care: Experience of a Brother of a Sick Child in Cameroon. In: Erhabor O, Munchi A, éditeurs. Human Blood Group Systems and Haemoglobinopathies. London: Intechopen. 2021; 472-478.
17. Mboussou M. Healing in the round: on the front of Sharing therapeutic space in black Africa. Rib. 2002; 1: 22-25.
18. Menick DM. Social and cultural perceptions of child disability in Sub-Saharan Africa. Perspect Psy. 2015; 54: 30-43.
19. Engolo CE. Socio-economic changes and living conditions of rural households in Cameroon. Rev Fr Sociol. 2001; 42: 281-294.
20. Scelles R. Fraternal bonds and disability: from childhood to adulthood, suffering and resources. Toulouse: Eres. 2010.
21. Bourhaba S. Singularity and multiplicity of sibling relationships, Abstract. Cah Crit Therapy Fam Prat Networks. 2004; 32: 23-33.
22. Ndjiengwe F. From physical stigma to symbolic marking: evolution of identity construction in sickle cell disease. In: Lainé A, Bonnet D, Keclard L, Romana M, editeurs. Sickle cell anemia Perspectives on an orphan disease. Paris: KARTHALA Editions. 2004; 221-228.