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Research Article

The Psychological Impact of Sickle Cell Disease on the Sick Child's Family

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Abstract

Background: Sickle cell disease is the most common genetic disease in the world. It manifests in children mainly through localized or generalized pain attacks on the body, chronic anemia and higher susceptibility to infections. Sickle cell crises are most often chronic, unpredictable and can lead to multiple hospitalizations. Because of its representation as a death sentence of the affected child or as a manifestation of the transgression of an ancestral norm by the family, sickle cell disease is a traumatism and a taboo in sub-Saharan African families where it is a forbidden subject between children and adults or even between children.

Objectives: This review provides a theme-by-theme review of international publications on the implications for family dynamics of the presence of a child with sickle cell disease.

Methods: Access to the publications was achieved through the exploration of 5 databases with the entries "Sickle Cell Disease, family" and "Drépanocytose, famille" being used for the search.

Results: We identified 25 research projects in Africa, Europe and America, published in English or French between 1988 and 2019, which consisted of general articles on the subject and the experience of practitioners working in hospitals with children suffering from sickle cell disease and their families. These researches emphasize the psychological suffering of the sick child as well as of his parents, siblings and the couple of his parents.

Conclusion: Professionals have to take into account the experience of family members through a psychosocial support mechanism adapted to the needs of the family group and each of its members without simply focusing on the child-mother relationship. This can make the family a resource for the sick child, family members and caregivers

Keywords: Sickle Cell Disease; Sick Child; Family; Psychological Experience; Literature Review

Introduction

It is accepted that a supportive and containing family environment conducive to a therapeutic alliance being established in the best conditions is a necessary condition for improving the quality of life of the sick child and his family [1,2].

Few studies focus specifically on the experience of families including a child with sickle cell disease. However, it remains a common pathology characterized by physical suffering, repeated hospitalization, side effects from the treatment and even risk of mortality.

This genetic disease is the most prevalent in the world, with about 500 million individuals carrying the sickle cell trait and 50 million living with the disease worldwide [3]. Originally spread over malaria-endemic areas such as sub-Saharan African countries, migratory flows have gradually changed its distribution worldwide; it is found in virtually every country having African origin populations [4].

With a prevalence rate of 2% in the general population [5] and an estimated death rate of more than 70% among children under 5 years of age [6], Africa remains the continent most affected by this disease. In France, Brazil, India, the USA and many other non-African countries, it is the first genetic disease for migrant patients from sub-Saharan Africa and the Mediterranean region.

This pathology discovered in 1910 in the USA is a pathology of hemoglobin handed down to the child by both parents. This is due to the presence in the blood of abnormal hemoglobin's, which cause a deficiency in oxygen supply to the body's various organs by the red blood cells. The rapid break down of red blood cells

and lack of oxygen lead to anemia and chronic pain [4,7]. This is a chronic disease-giving rise to multiple hospitalization incidents, unpredictable crises and great suffering with potentially lethal outcomes and expensive treatment [8].

This general review provides a review of the literature that takes stock of the state of humanities and social sciences studies on the impact of sickle cell disease on family dynamics and on each of its members. These studies were conducted out in the patients' countries of origin in Africa and in migration situations in Europe and America.

The results are presented in relation to: 1) the methodologies adopted; 2) the populations concerned; 3) a summary of the results of the studies analyzed. We end with a discussion of the results.

Materials and Methods

Exploring 5 databases (Cairn Info, Direct Science, MEDLINE, PubMed and CINHAL with keywords "Sickle Cell Disease, family" or “Drépanocytose, famille”) we obtained 42 publications. 17 of these were excluded from the analysis due to their purely medical orientation. 25 works appearing between 1987 and 2018 were thus analyzed. Looking at these articles’ bibliographies, 6 chapters of publications addressing the theme were identified.

Only the articles, which are interested in the experience of the members of the family in a qualitative and not quantitative approach, were included in the analysis. Thus, the results of this review are not quantified; they highlight the psychological processes at work within the family and at each of its members. The effects are not quantifies with validated scales. The selected studies are qualitative or qualitative and quantitative studies.

Each publication was analyzed according to the following criteria:

- Place of data collection; authors’ specializations
- Population concerned; methodology
- Main results.

The researches were conducted in Africa, Europa and America. France is the country in which the most research was conducted (Table 1). Different specialists conducted them either individually or within a research group, more in single-discipline teams than in multidisciplinary teams with a predominance of teams composed of physicians (Table 2). Various populations were concerned by the researches that focused more on the experience of the sick child and his parents (the mother in particular) (Table 3).

Only 3/24 researches [9-11] met the participants in their homes. All the other research was conducted in care institutions for sick children. The majority of research thus gives little interest to the experiences of families and sick children outside hospitals.

Global description of research

Almost all continents are represented by the research cited in this work. This gives a universal overview of the impact of sickle cell disease on families without, however, denying the existence of specificities linked to countries, societies, families and individuals in the experience of sickle cell disease.

Places		Number of publications
Africa	Cameroon	4
	Nigeria	1
	Niger	1
	Democratic Republic of Congo	1
	Togo	1
Europe/ Africa	France/Côte d’Ivoire	1
Europe	France	10
America	USA	5
	Brazil	1

Table 1: Places of research.

France, USA and Cameroon are the countries are the countries in which a lot of research has been done concerning the psychological impact of sickle cell anemia on the family.

Anthropologists, doctors, social workers and psychologists, carried out the research individually or in research groups.

Specialties			Number of publications
Individual [6]		Medical doctor	1
		Anthropologist	2
		Psychologist	3
Research group [19]	Multidisciplinary [7]	Nurses and medical doctors	1
		Psychologists and medical doctors	2
		Psychologists and social workers	2
		Psychologists, medical doctors and anthropologists	2
	Monodisciplinary [12]	Psychologists	5
		Medical doctors	7

Table 2: Authors’ specializations.

Very few researchers have worked alone. Collaboration with colleagues seems to be a resource for most researchers. Most of the studies have been done in research groups. Monotidisciplinary research groups are the most invested by researchers. As a result, researchers would feel more comfortable collaborating with colleagues in the same discipline. Research done in multidisciplinary

groups represents less than half of the research done in research groups. This may reflect, despite the recognition of the interest of working with colleagues from other disciplines and taking into account the global dimension of patients and their families, the difficulty researchers face in collaborating with colleagues other specialties.

The parents, the sick child and his brothers and sisters are the main research population.

Populations	Locations	Number of publications
Sick children and adolescents aged 9 to 16 [4]	Cameroon	3
	France	2
Sick children and their parents [11]	France	7
	USA	3
	Brazil	1
	Democratic Republic of Congo	1
Parents of sick children [7]	Nigeria	1
	Togo	1
	Niger	1
	Cameroon	1
	France	1
	USA	2
Nursing staff [1]	Ivory Coast and France	1
Parents and siblings of sick children [2]	USA	1
	Cameroon	1

Table 3: Research populations.

Most of the research has focused on the psychological experience of sick children and their parents in general, and of mothers in particular. Only two studies were interested in the psychological experience of parents and siblings of the sick child. Researchers pay little attention to the psychological experience of brothers and sisters of sick children. There is not taking into account the psychological experience of members of the extended family of the sick child (grandparents, cousins, nieces, uncles ...). This contrasts with the fact that sickle cell disease is thought of as an illness of the family, which often gives rise to the mobilization of the various members of the extended family to find a solution to this evil, which concerns them all.

Researchers used Case report, literature review, clinical interview, drawing, scales and observation are the data collection tools either exclusively or in a complementary manner.

Clinical interviews is the most used data collection tools, exclusively, by researchers. It is followed by the questionnaire used more in a complementary way with the drawing than with the questionnaire. Researchers as investigative tools also use case studies and the literature review. The use of psychometric scales is not well used.

Protocols		Number of publications
Clinical interviews		9
Questionnaire		4
Interview and questionnaire		2
Interviews and drawings		4
Case report		3
Literature reviews		2
Psychometric scales	Kidcope inventory	1
	Children's self-efficacy for peer intervention scale	
	Social support scale for children	
	Coping health inventory for parents	
	Family relations scale	
	Child behavior checklist	

Table 4: Data Collection Protocols.

The parental couple

The parents play different roles in caring for the sick child. The father is seen to pay for the child's medical benefits while the mother looks after organizing the care itself [12,13]. This disease's family economy is part of the African cultural tradition which makes the mother responsible for the health and education of children [6].

Alteration of conjugal relations

The sick child's presence in the family calls into question the legitimacy of the marriage by the parents [14] and the extended family's members [6]. This a source of guilt for the parents.

Some authors instance cases where the parents blame each other for handing down the disease to the child. Often, the mother is accused by the restricted and enlarged family as having brought "bad luck" and finds herself facing divorce [12,15].

Thus, due to the pressure exerted on the couple by the extended family (especially that of the male partner and sometimes with his connivance), a deterioration in the parents' conjugal relations can be observed as they fail to afford mutual support in the crisis situations experienced with their child [6,13,15]. According to these authors, the ensuing climate of conflict constitutes, alongside economic difficulties, a major factor in the breakdown of conjugal ties.

The presence of a sick child changes family habits, related to the daily care required by the sick child, to his multiple hospital-

izations, the search for an appropriate diet to cater for his health needs and the precautions adopted to prevent and control his crises [16].

The difficulties encountered in caring for the sick child are multiple 1) fear of having another sick child, 2) uncertainty as to the future and survival of the sick child, and 3) pressure from the male partner's family. Indeed, divorce can be perceived by both parents as providing the solution to put an end to the curse [17].

Resistance of the parents to social and family pressures

Some couples manage to resist pressure and develop strategies of mutual support [12,15] note that in this case, the couple may represent a source of support for the mother, regardless of her educational level and socio-professional status. They emphasize the socio-familial's isolation and denigration of couples who resist social and family pressures.

To avoid this marginalization, parents hide the existence of their child's illness: 1) to save their union and 2) to prevent stigmatization and violence against the sick child from some members of the family and society [6,17].

Effects on the mother

Most of the authors included in the study focused essentially on the mother's experience.

Psychological suffering

Mothers suffer greatly from the distress of their sick children [6,18] note that the frequency of child seizures and the extent of social limitations caused by their illness influence mothers' stress. They note the existence at home of psychological suffering such as stress, irritability and depression. Stress becomes exacerbated during seizures and during hospitalizations of the sick child, periods during which the mothers witness his suffering [13].

The mothers' social isolation is related to their uncertainty as to the onset of crises and their overinvestment in care of their sick children [1-3]. Withdrawal from socio professional activities is accompanied by a tendency for mothers to neglect their own needs and personal care [13].

Early death and anticipatory grief

Parents are always drawn into anticipatory bereavement for their sick child, symbolizing apprehension of their own death [12]. They perceive the diagnosis of sickle cell disease as an announcement of death and the ensuing crises reactivate their fears about the death of the sick child [4,20].

The sick child's death undermines the mother's social status and narcissism [6]. In sub-Saharan African culture, a sick child remains a source of valorization of maternal narcissism since the woman's social status is identified with her ability to have children and take care of them. According to [11], this explains parents' refusal to contemplate abortion in the case of prenatal diagnosis of sickle cell disease. He emphasizes that parents remain extremely pessimistic as to how they see their child's future, while remaining attached to a hope of healing, based on their magico-religious beliefs.

Treatment of the sick child

[21] explains the verbal and/or physical aggression of mothers against their sick children during hospitalizations in France as an unconscious manifestation of their wish to see them die. Meanwhile [17] consider that through this aggression, mothers seek to strengthen their sick child's resilience against the unpredictable violence of the disease.

Guilt

Non-African researchers situate the suffering of mothers on the axis of maternal guilt that has several origins: 1) medical discourse on the disease's transmission; 2) cultural representations of the disease; 3) the mother's helplessness in looking after her child and sparing her suffering and crises [4,12,13,20]. The research places particular emphasis on the fact that society attributes to the mother the responsibility for the transmission of the disease to the child and leads her to internalize this guilt that is not mentioned concerning the fathers.

African researchers working in Africa set the mothers' suffering against the background of the group dynamics. They are seen to refuse being penalized for a disease for which they are not responsible. They suffer from the indifference of society, the state and the family to their situation and that of their sick child [6,15,17,22].

The experience of the fathers remains an enigma. African and non-African researchers tend to focus more on the experiences of mothers. This interest for the mother can be understood by its accessibility because it is she who is most often at the bedside of the sick child in the hospital. Since research is generally conducted in care institutions for the sick child, it is obvious that the mother is the family member most accessible to researchers.

Offspring of the sick child and the desire to have children

Mothers suffer from their understanding that their sick children's medical treatment could make them infertile. Indeed, they may oppose any treatment that would result in their sick child's sterility, be they a girl or a boy [4,14,23]. They would like their sick children to perpetuate the family by, in turn, making children.

There is a diminished desire to have further children in mothers who are afraid of having another sick child and having to again endure the suffering their care entails. However [6] emphasizes that none of them relinquish the desire to have other children. He explains this for 3 reasons; 1) even a sick child is always a source

of gratification, narcissism and social esteem for a mother; 2) the random nature of sickle cell disease transmission; 3) the desire for their child to be part of the succession of generations.

Effects on parent/child relationships

The effects of sickle cell disease on relations between parents and children are manifold.

Sickle cell disease, a family taboo

Parents conceal the name of the disease from children or at least from some of them while children also hide their knowledge of the disease from their parents [12]. Parents seek to protect themselves and their children from stigma and the idea of death and children want to protect their parents.

Overprotection of the sick child

Parents tend to overprotect their sick child [11,13,17]. This results in continuous monitoring of his state of health, restricting his physical activities and imposing less educational constraints. For these authors, this can lead the child to become capricious and bad tempered while the educational advice given to parents on the matter remains ineffective.

This protection constitutes a counter-investment of the parents' guilt and anger at the existence of this illness and their inability to heal their child [11,17]. It is therefore useful to support the psychic work of the mothers with this overprotection in order to enable them to transform it and not to make it an obstacle to the autonomy of the affected child.

Effects on families of relations with the medical community

In most sub-Saharan African countries, families bring their children to hospital only in the event of serious crises, numerous failures of self-medication [15,22] and following several therapeutic remedies supported by members of the extended family. The latter, although stigmatizing the family can become major helpers by devoting time and money in efforts to heal the child [4].

Due to the coexistence of traditional and modern representations of sickle cell disease [4,10], families often hide the existence of a combination of traditional and modern care from the caregivers [10,21,24]. They generally show resistance to certain medical practices such as transfusion by the fear of contaminated blood and by the child becoming possessed, through this vital fluid, by some unknown and potentially malevolent being [4,12,25].

Effects on the siblings of the sick child

The experiences of siblings of the sick child are generally evoked indirectly through observations made by children with sickle cell disease or their parents [12,13,15-17]. Only [9] and [10] involved siblings of the child with sickle cell disease.

Jealousy and guilt

The overprotection of the sick child leads to parents gradually depriving the other children of care [22]. The latter feel abandoned, marginalizes and excluded by the parents and the extended family [10,17]. They express jealousy towards the patient whom they identify as the main culprit for the crises that destabilize the family financially and emotionally [17].

[10,16] point out that the siblings of the sick child feel guilty for having negative thoughts such as jealousy against the latter. According to these authors, they consider themselves «bad brothers/sisters".

The brothers and sisters express their helplessness to heal the sick child, to foresee his crises, to satisfy the parents, to make that the sick brother has a place like the others in family and outside [10]. They suffer from their inability to predict crises and their consequences both on the patient and on the parents as well as on themselves. This helplessness generates feelings of guilt about the sickness of the disease and the sick brother [10] argue that parents and professionals should be more aware and understand the difficulties faced by siblings.

Parenting by the sick child's siblings

Their parents to assume a parental role in watching over and caring for the sick [11,12] whether older or younger than the sick child, often enroll siblings. The siblings of the sick child are therefore a resource for their parents regarding their surveillance and protection against crises. Siblings describe the sick child as passive a passive victim of the desease within the family.

There is a very high level of involvement of the sisters of sick children in their follow-up [24]. This involvement is due to the educational model of girls in African families that aims to make them mothers, people who can take care of children.

The question of death concerning the child reaches

The children evoke the fact that their sick brother could die with the idea that this death might not make the disease disappear. Indeed, if their brother dies, then the disease could reach them. This "transfer" of the disease can occur when the child is dying or healing. However, since healing is not envisaged, only death leads to an "other" child incarnating this evil.

Children express the desire to die for their sick brother or sister who destabilizes the family and incurs expenses for their care. They also want the death of this child who, during his crises, becomes strange. This desire is based both on the level of reality (expenses related to care, fear generated by crises) and fantasies (it will not survive) [10]. These children therefore oscillate between the desire and the fear of the death of their sick brother or sister

who, despite his illness, remains "other", the same; to desire one's death is, finally, to evoke one's own death in a confusing process of identification/differentiation usually in the context of siblings.

The quality of parent's relationships can predict the adaptations of siblings on the sick child disease. The family processes such as coping and mutual support between parents reinforce the psychological adjustment of the sick child's siblings whereas family conflicts predispose them to a poor psychological adjustment to the constraints involved [9].

Effects on the sick child

The sick child's relationships with family members are impacted by his illness.

Fragility of the family psychological envelope in a hospitalization situation

When children are hospitalized in Africa, the family stays with them. In France, when they are hospitalized, the family cannot stay alongside them; the caregiver takes a central role in ensuring the child's psychic well-being [25].

[7] insist that sick children feel abandoned by their family during their hospitalizations in France. According to them, they invest in the caregivers who provide a base of emotional security for them. These authors refer to a "medical psychic envelope" to account for the psycho-affective security that medical staff represent for hospitalized children. In their view, during these long hospital stays far from home, the sick child feels more protected by his medical psychic envelope than by his family and cultural psychic envelope. They specify that this psychic reality is generally observed in sick children hospitalized for a relatively long period as in the case of stem cell transplant operations that usually take place between 8 and 11 years of age.

The many hospitalizations the sick child undergoes weaken his body psychic envelope and his family psychic envelope [7,25]. Hospitalizations are experienced by sick children as a "third party separator" intruding into their relationships with family members [26].

Children are often hospitalized following fits of pain or chronic anemia, which are the main symptom of sickle cell anemia, which remains a taboo disease in families [27] studied, through interviews and drawings, the impact of the family taboo concerning sickle cell anemia on the structure of the body image of sick children. There is a complex link between knowing about but not understanding their disease, and showing/saying something of their body, as they experience it. The drawing shows both the need and desire to discuss and express their pain, and the fear of doing so, as well as the difficulty of finding the words to express and represent the strange familiarity that is their disease. The family taboo around the disease is an obstacle to the construction of body image in the children, who have shown difficulties in talking about their body and their disease with their family members.

Emotional rapprochement with the spinal cord donor

Rapid and substantial progress in genome editing approaches have proven valuable as a curative option given plausibility to either correct the underlying mutation in patient-derived hematopoietic stem/progenitor cells, induce fetal hemoglobin expression to circumvent sickling of red blood cells, or create corrected induced pluripotent stem cells among other approaches [28]. Nowadays, bone marrow transplant from a brother or sister with HLA compatibility cures sickle cell disease in about 95% of cases [3]. This therapy, as well as gene therapy, is absent in some parts of the world, such as African countries were sickle cell disease sickle cell disease remains synonymous with the death of the sick child.

[7] remark that having a bone marrow transplant in France elicits a stronger affective relationship with the donor than with the other siblings. They consider this rapprochement as a means used by the sick child to meet his debt towards the donor of the "healthy marrow".

Feeling of rejection and stigma

[29] presents a case of a sick teenager suffering from the mockery of his entourage on the body marks left by the disease as with the yellowish color of her eyes. She is a victim among her siblings of many frustrations and restrictions following her illness, being excluded from certain games.

Sick children cope with repeated onsets of pain [3] which cause them to experience feelings of distress and loneliness among their siblings compounded by their difficulties to share this torment with their loved ones [26]. They also cope with their verbal abuse by some family members who sometimes describe them as "children of death" who ruin their families before dying [12]. This family reality awakens in them an anguish of death and confronts them with deadly fantasies and an elusive fear [26].

Sick children feel very little support from their non-sick siblings. The latter overprotect them against crises and behave with them as if they were passive victims of the disease, children to be protected. Therefore, they do not give them space and time to say and express the way in which they experience their illness. This leads them to feel more isolated in the family and not supported by siblings [27].

Guilt

The sick child feels guilty for the family's dysfunctions resulting from the economic difficulties it encounters in providing him with care and overprotection [7,25].

[30] interpret the desire to find employment, expressed by some sick adolescents as a mechanism to pay back their debt to their families by helping their parents finance the care afforded.

Projection into the future

Sick children remain frozen in a present marked by excruciating pain with which they identify and direct almost all their psychic energy towards their own pain-wracked body and divest from the other objects in their environment [3].

For adolescents, parenting's questions take a special interest. They are keen to pair up with a spouse who does not carry the gene for sickle cell disease [14].

Abuse of the sick child

Only 2 publications [29,30] deal with ill-treatment of the patient. In both cases, the authors work in child protection agencies, giving them access to situations of abuse. It appears that this situation is difficult to observe outside clinical case studies.

[30] present cases of abuse of children with sickle cell disease by their family members in Cameroon. They describe a state of dysfunctional parenthood caused by the socioeconomic constraints of caring for those children. They state that, because of the parents' inability to finance both medical care and schooling, sick children often quit school to continue to receive medical care. They specify these children, being stigmatized as children of misfortune, suffer from both physical (physical abuse and neglect of body care) and psychological (insults, mockery) ordeals. Indeed, some run away from home while others have a renewed desire to drop out of school and to get a job in order to support their parents financially in managing their illness [30].

Discussion

The results differed depending on whether the studies take place with migrants or with families living in their country of origin. While the authors all evoke the guilt, the affects raised by desires and the anguish of death, not all ascribed the same interpretation to these affects, with the inferences drawn seeming to be influenced by the authors' cultural and theoretical backgrounds. These results are to be understood and construed taking into account the transferential phenomena between the authors and their participants in the sense of Devereux [31].

The experience of the sick children themselves, of their siblings and of the father were virtually absent from the publications. The centralization of studies on the mother's experience is, in part, because data were collected at the hospital and that mothers stay with their children or accompany them there. However, this does not explain the few works focus on what the sick child experiences through within the family.

As far as the child is concerned, it would be interesting to understand his investment in the caregivers during long periods of hospitalization. It would also be interesting how he manages potential conflicts of loyalty between caregivers and parents. A better understanding of family and caregiver's relationship would certainly help prevent needless suffering. This also implies taking into account representations of the disease for all concerned.

There is an absence of studies on the palliative care of sickle cell patients. This may shed light on the difficulty of caregivers in predicting the death of these patients despite the severity and spontaneity of the crises, even if these crises often return, for families, like the imminent death of the patient [27].

Almost all of these studies concerned African families or families of African descent in France, in Brazil and in the USA. It would be interesting to extend the study to Asian families or families from Asia (India, Patkistan, Bangladesh…) affected by sickle cell anemia.

Conclusion

This review presented elements evoked about other situations of illness [32-36] such as guilt, fear of death and the desire to have children. It showed the need for further research to better understanding the impact of the disease on family dynamics by cross-referencing data from different methodologies and countries.

It also showed the widely varying impacts the disease has on family dynamics and on each member, that merit further multidisciplinary study to identify what appears to enhance or disrupt family life with a child affected by sickle cell disease.

Studies highlighted the major impact the disease has on the family as a group and specifically on each of its members whether they are in migration situation or not. This strongly suggests the need, during hospitalization of the sick child, to take into account the experience of family members through a psychosocial support mechanism adapted to the needs of the family group and each of its members (siblings, father, mother, grandparents, uncle, aunt…) without simply focusing on the child-mother relationships.

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Conflict of Interest

The authors declared no conflict of interest.

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