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(RESEARCH ARTICLE)

Problems of parental care for sickle cell children at the ''Gracia Fondation'' treatment center

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Abstract

Introduction: Sickle cell disease is a complex disease, both historically and medically, at the collective and individual levels. It is the world's most common genetic disease, affecting around 50 million people, and its treatment is multidisciplinary, comprising 2 components: analgesic treatment and treatment of factors promoting sickle cell disease. The sole objective of this study is to identify the various problems related to the care of sickle cell children encountered by parents at the ''Gracia fondation'' Center.

Methodology: This is a descriptive, cross-sectional and quantitative study whose population is made up of 20 parents of sickle-cell children followed up at the ''Gracia fondation'' center in the city of Kisangani from 15 /10 to 15 /11 2024. We used an interview technique to collect data, based on a pre-established questionnaire.

Results: After tabulation and analysis of the data, the following result was observed: insufficient financial means and abandonment of children by their fathers are the main difficulties experienced by parents in caring for their sickle-cell-affected children, according to 100% and 80% of the respective subjects.

Keywords: Problem; Care; Child; Sickle Cell; Parent.

1. Introduction

According to the International Covenant on Economic, Social and Cultural Rights, States Parties to the Covenant must take “the necessary steps to ensure the reduction of the stillbirth-rate and of infant mortality, and the healthy development of the child” (Article 12). Sickle-cell anemia is a genetic disease that can be described as global, and which seriously affects children's health, even leading to their death. This transcontinental and trans-ethnic disease has been forgotten by the international community, yet it was recognized as a public health priority by the WHO in 2005 and by the UN in 2008 (Resolution A/63/237) [27].

Sickle cell anemia is a complex disease, both historically and medically, at the collective and individual levels. It is the most common genetic disease worldwide, affecting around 50 million people [1].

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Hemoglobin abnormalities caused by point mutations in a heme chain lead to red blood cell falciformation in stressful situations (infections, hypoxemia, acidosis), resulting in occlusion of blood capillaries and particularly painful bone infarctions, which explain the vaso-occlusive crises that bring patients to the emergency room [2].

The structural anomaly in hemoglobin is represented by a DNA point mutation characterized by the substitution of an amino acid, glutamic acid, by valine, in position 6 of the θ chain. The anomaly is located on chromosome 11. Genetic transmission is autosomal recessive. When only one chromosome carries the HbS gene (transmitted by the mother or father), the subject is said to be heterozygous; a healthy carrier. However, when both chromosomes are carriers of the gene (transmitted by the mother and father), the subject is said to be homozygous and the carrier is ill [4].

Depending on the genotype of the parents, the following combinations are possible: 1. AA children born to parents who are both AA, who are not ill and cannot transmit sickle cell disease to their offspring; 2. AA and AS children, if one parent is AA and the other AS. AS children are not sick, but can transmit sickle cell disease if they marry another AS or SS person; 3. AA, AS and/or SS children, if both parents are AS and AS; 4. SS children if both parents are SS and SS [1].

Until the 1990s, sickle-cell anemia was considered a fatal disease, with studies reporting a life expectancy of no more than 48 years even in developed countries. However, major advances in our understanding of the still highly complex pathophysiology of the disease, and the growing codification of its management, have made it possible to reduce mortality considerably by adhering to certain simple rules, and to transform sickle cell disease from a “fatal disease” into a “chronic disease” [3].

Sickle cell anemia is a genetic disease, inherited from both parents. It is the most widespread genetic disease in the world, affecting over 50 million people, with a predominance in Africa, India, the Mediterranean Basin and the Middle East. From these regions, it has spread to other countries as a result of population movements migration [5].

In France, sickle cell anemia is the most common genetic disease, affecting 20,000 patients by 2020, making it the leading genetic disease ahead of cystic fibrosis. However, over 50% of sickle cell patients live in the Île-de-France region [6].

Sub-Saharan Africa is particularly hard hit: one in every 65 newborns there has sickle cell disease. sickle cell disease. In Mali, the prevalence of sickle cell disease is estimated at 12%, including 1-3% for the homozygous form, making it a public health problem due to its frequency and multiple complications [7].

In Burundi, the prevalence of sickle cell disease is unknown. In 2016, the WHO estimated that it was at least 2% and that 10% to 40% of the Burundian population were carriers of the sickle cell gene [1].

Sickle cell disease presents as hemolytic anemia interspersed with vaso-occlusive crises, often complicated by severe bacterial infections. Pallor and subicterus, indicative of chronic hemolysis, are almost constant. The clinical signs of sickle cell disease are synonymous with complications. Sickle cell disease is manifested both by noisy acute manifestations and silent chronic complications: splenic sequestration crises, hyperhaemolytic crises, aplastic crises, osteo-articular vaso-occlusive crises or bone infarcts, extremity syndrome (hand-foot syndrome), cerebral crisis due to cerebrovascular accident (CVA), abdominal crisis or diffuse abdominal pain, priapism and clitorism, hypersplenism or splenomegaly, growth retardation and delayed puberty, severe infectious episodes, etc. [26].

In the Democratic Republic of Congo, sickle-cell anemia is a serious chronic disease responsible for early mortality and reducing personal, social and family quality of life, and family life. Patients may be experiencing great social difficulties, family conflicts, professional problems (precarious work and sometimes unsuitability for the job and repeated absences) or school problems (repeated absences, misunderstanding of classmates and the school system), and are therefore likely to have a greater number of attacks [8].

The main factors triggering vaso-occlusive attacks are cold, altitude (air travel), stress (school or university exams, social life, etc.), infections and dehydration.

Sickle cell vaso-occlusive crisis is not usually accompanied by a major drop in hemoglobin levels. The occurrence of acute anemia in the course of sickle cell disease should raise the possibility of central complications such as bone marrow necrosis, parvovirus B19 infection and acute folate deficiency, or peripheral complications such as acute splenic sequestration and immunological transfusion accidents. This crisis is the most frequent acute complication of sickle cell disease, with an estimated prevalence of 95%. The painful vaso-occlusive crisis may sometimes herald another, more serious complication, or conceal other differential diagnoses [9].

Spontaneous mortality from sickle cell stroke can be as high as 20%, reflecting the extreme severity of this complication. The relative risk of developing a stroke in sickle cell disease is much higher in children than in adults, with a predilection for strokes between the ages of 2 and 10, and a peak after the age of 30 [10].

Therapeutic management is multidisciplinary, with internists, emergency physicians and resuscitators all involved in the management of sickle cell complications. It includes symptomatic treatment of vaso-occlusive crises, transfusion, or even exchange transfusion, and organic support depending on the severity of the crisis. Patients with severe complications are admitted to intensive care [7].

The treatment of a simple vaso-occlusive crisis comprises 2 components: analgesic treatment and treatment of factors favoring falciformation. The vaso-occlusive crisis is the cause of very intense bone pain at a level superior to that of a bone fracture. The therapeutic response must therefore be at the same level, and in all cases requires major analgesics such as morphine [11].

According to the African Society of Pediatrics (2018), in sickle cell disease, there are only palliative treatments. The bulk of treatment consists of managing the symptoms resulting from crises. Sickle cell patients need rest, good oxygen therapy in the case of thoracic syndromes, good hydration in hot weather, and analgesics in the case of pain. Emergency transfusion remains the only treatment for profound anemia. Folic acid is given for life to facilitate cell renewal [12].

Around 40% of adult sickle-cell patients present with intermittent or acute priapism at some stage in the course of the disease. Attacks are most often nocturnal and favoured by the presence of sleep apnoea syndrome. The major functional risk is definitive fibrosis of the corpus cavernosum. Systematic anamnestic screening and management of intermittent priapism are essential to prevent the onset of acute priapism, often with permanent sequelae [11].

Any sickle cell complication should a priori be treated in hospital, due to the unpredictable risk of aggravation or association with other conditions that may have been masked at the outset: any bone pain crisis that is abnormal in terms of duration, intensity, resistance to usual initial treatments, fragility of the patient's underlying condition, or the impossibility of providing adequate analgesic treatment and hydration on an outpatient basis, should be referred to hospital. Furthermore, all extra-osseous complications are systematic indications for hospitalization, with hyperthermia leading the way, as well as thoracoabdominal pain and vaso-occlusive syndromes, and severe anemia. The efficacy of curative management of acute complications of sickle cell disease depends on the quality of initial management of vital distress and proper diagnostic evaluation of patients [13].

The criteria for returning home during a vaso-occlusive crisis are absence of fever, absence of chest pain, Respiratory Rate below 20 movements per minute, no morphine injections for more than 8 hours [14].

Sickle-cell anemia can lead to the death of the child, especially when the disease is not detected and treated from birth, as is the case in many African and developing countries such as the Democratic Republic of Congo.

In most countries where sickle cell disease is a major public health problem, there are no national programs to combat it. The basic structures needed to care for patients are generally lacking. Systematic screening for sickle cell disease is a simple blood test, yet this is not common practice. In developing countries, the disease is usually diagnosed only when a serious complication occurs. As a result, over 50% of children with the most severe form of the disease die before the age of five, most often from infection or severe anemia. Early detection of the disease is essential, so that couples can benefit from information, education and appropriate care [15].

Effective ways of improving the health of sickle-cell children include: 1. standard treatments: antibiotics, vaccines, folates; 2. treatment of vaso-occlusive crises: analgesics and oxygen therapy; 3. prevention of factors triggering crises (cold, altitude, infections, dehydration); 4. Take folate-rich medication for anemia; 5. Vaccination to prevent pneumococcal and meningococcal infections; 6. Blood transfusion in cases of severe anemia or infection; 7. Blood transfusion to reduce the proportion of hemoglobin S [16].

Sickle cell disease is one of the most common diseases in the Democratic Republic of Congo, as recent epidemiological data demonstrate. They show that 2% of newborns are homozygous for hemoglobin S, and that an estimated 40,000 sickle-cell children are born each year, half of whom die before the age of 5... although early and appropriate treatment enables patients to control their symptoms and avoid serious crises [17].

Sickle-cell anemia is linked to the representation of death in the African context. Families live in uncertainty in the face of this tragedy. Faced with this situation, sickle cell disease remains hidden from the family circle; only the parents

manage the situation, even during the child's vaso-occlusive crisis. Given the sensitivity of the information we were seeking, a quantitative approach was used to highlight the problems associated with the management of sickle cell disease in our study environment. This approach is facilitated by the community diagnosis process, which enables a community to identify its own problems and needs, and the resources available to address them.

In view of the above, we pose the following research question:

What are the various problems related to the care of sickle-cell children encountered by parents at the ''Gracia fondation'' Center?

1.1. Objective

Our sole objective is to identify the various problems encountered by parents of sickle-cell-affected children at the ''Gracia fondation'' center.

1.2. Aim of the study

With a view to improving the health of sickle-cell-affected children in the Democratic Republic of the Congo, this study has two interests:

Scientific and educational: this study is intended to make a scientific contribution in terms of new knowledge, as a library that can help researchers in their future studies.

On a practical level: this research is intended to be a frame of reference, raising awareness both for administrative and health authorities to think about improving the care of sickle-cell children, and for parents to bring their children to specialized centers for proper care.

The results of this study will enable us to formulate a number of concrete actions (possible solutions) that could help decision-makers to draw up and implement national programs to combat sickle cell disease throughout the Republic.

2. Methodology

2.1. Type of study

This is a descriptive, cross-sectional and quantitative study on the problem of caring for sickle cell children encountered by parents at the “Gracia fondation” treatment center in the town of Kisangani, in the Democratic Republic of the Congo, from 15/10 to 15/11 2024.

2.2. Study population

The population of this study is composed of parents of children with sickle cell disease followed at the ''Gracia fondation'' Sickle Cell Treatment Center in the city of Kisangani during the period from 15 /10 to 15 /11 2024.

Given the importance of this study, our sample included 20 subjects representing the parents of children who were followed up during this period.

2.3. Data collection methods and techniques

In view of all these considerations, and taking into account the nature and purpose of our study, as well as the different levels of education of parents of children with sickle cell disease, we opted for an interrogatory method based on a questionnaire that we developed and administered or submitted to the respondents, in the form of a structured interview to obtain information on the difficulties associated with caring for children with sickle cell disease.

After finishing with the administration of the ''Gracia fondation'' center, we were accompanied by a nurse to collect data from the parents. Due to time constraints, data collection took place every Tuesday and Friday from 10am to 2pm.

2.4. Data processing

The data collected were analysed and encoded using Microsoft Office Excel 2013 and SPSS 20.1. The results obtained were presented in the form of tables. Data analysis was made possible by calculating frequencies and percentages.

3. Results

3.1. Socio-demographic data

3.1.1. Gender

Table 1 Gender distribution of study subjects

Gender	f	%
Female	13	65
Male	7	35
Total	20	100

Analysis of Table I shows that 65% of study subjects were female, compared with 35% male.

3.1.2. Age

Table 2 Age distribution of study subjects

Age range in year	f	%
20 - 29	5	25
30 - 39	8	40
40 - 49	4	20
50 and over	3	15
Total	20	100

Analysis of Table II shows that the study population is made up more of subjects in the 20 to 39 age bracket, i.e. 61.1%, while those aged 50 and over represent only 15% of cases.

3.1.3. Level of education

Table 3 Distribution of respondents by level of education.

Level of education	f	%
University	8	40
Secondary	12	60
Total	20	100

For the “level of education” variable, analysis of Table III shows that the majority of study subjects were at secondary school level (60%), while those at university level accounted for only 40%.

3.1.4. Marital status

Table 4 Distribution of respondents by marital status.

Marital status	f	%
Married	14	70
Single	6	30
Total	20	100

This study revealed that 14 subjects, or 70%, were married, compared with 30% who were single.

3.1.5. Socio-economic level

Table 5 Distribution of respondents according to socio-economic level.

Socio-economic level	f	%
Low	11	55
Medium	7	35
High	2	10
Total	20	100

The data in Table V show that 55% of the subjects in the study had a low socio-economic standard of living, while the socio-economically average represented 35% of cases. On the other hand, two subjects (10%) were of high socio-economic status.

3.2. Variables studied

3.2.1. Circumstances of discovery of sickle cell disease in children

Table 6 Distribution of subjects according to circumstances of discovery of sickle cell disease

Circumstance of discovery of sickle cell disease	*f*	%
After child's second blood transfusion	8	40
Child's sickly state	6	30
Following pronounced pallor	4	20
Following growth retardation	2	10
Total	20	100

In Kisangani, episodes of blood transfusion and a sickly child are the circumstances in which sickle cell disease is discovered in 40% and 30% of cases respectively.

3.2.2. Number of vaso-occlusive crises per year

Table 7 Distribution of subjects by number of vaso-occlusive crises per year

Number of attacks per year	f	%
1 to 4 times	2	10
5 to 8 times	5	25
Several times	13	65
Total	20	100

The data in Table VI show that 65% of subjects report several vaso-occlusive attacks per year, while those who have 1 to 4 attacks represent 10% of cases.

3.2.3. Cause of vaso-occlusive attacks

Table 8 Distribution of subjects by number of vaso-occlusive attacks per year n=20

Cause of attacks	f	%
Malaria, diarrhea and typhoid fever	10	50

Cold and heat	7	35
Pregnancy if adult	4	20
Undecided	2	10

The data in Table VIII show that infections (malaria, diarrhoea and typhoid fever) and climate (cold and heat) are the main causes of vaso-occlusion in 50% and 35% of subjects respectively.

3.2.4. Management of children with sickle cell disease

Table 9 Distribution of subjects by elements of care for children with sickle cell disease n=20

Management of sickle-cell anemia	**f**	**%**
Pain control [treatment with analgesics (morphine)]	20	100
Transfusion in case of anemia	17	85
Ensuring good nutrition	15	75
Infection control (treatment with antibiotics)	14	70
Application of lifestyle hygiene measures	9	45
Hydration and oxygen therapy	7	35
Treatment with hydroxyurea	5	25
Treatment of priapism	5	25

Pain control, transfusion in case of anemia, good nutrition and infection control are the main components of sickle cell case management, according to 100%, 85%, 75% and 70% of cases.

3.2.5. Difficulties experienced by parents in caring for sickle-cell-affected children

Table 10 Distribution of subjects according to the different difficulties experienced by parents in caring for sickle-cell children n=20

Difficulties experienced by parents	**f**	**%**
Parents' lack of financial means	20	100
Abandonment of children by fathers	16	80
Socio-cultural problems	4	20

Insufficient financial means and abandonment of children by their fathers are the main difficulties experienced by parents in caring for their sickle-cell-affected children, according to 100% and 80% of the respective subjects.

3.2.6. Proposed solutions to improve sickle cell disease management

Table 11 Distribution of subjects according to proposed solutions to improve the management of sickle cell disease n=20

Proposed solutions for improving sickle cell disease	***f***	***%***
Government support for children with sickle cell disease	20	100
Separation of A/S and A/S or A/S and S/S couples	13	65
Sterilization of couples to avoid procreation	6	30

Government treatment of sickle-cell children by setting up national sickle-cell treatment centers throughout the Republic (100% of cases) and separation of A/S and A/S or A/S and S/S couples (65% of cases) are the solutions to improve care. However, 30% of subjects reported sterilization of couples to avoid procreation of sickle cell children.

4. Discussion and comments

4.1. Gender

Analysis of the study data shows that 65% of the subjects were female, compared with 35% male.

This result can be explained by the fact that it is women, mothers in most cases, who care for and accompany children to hospital for treatment. They are the primary family nurses, while the men are the ones who provide for their families.

4.2. Age range

The study indicates that the study population is made up more of subjects in the 20 to 39 age bracket, i.e. 61.1%, while those aged 50 and over represent only 15% of cases.

Given that the study population is made up of parents of sickle-cell-affected children, the 20 to 39 age bracket is therefore the age of majority. Both men and women can marry, thus founding their homes. These are the ages with which young people want to identify and confirm themselves in life.

4.3. Level of education

For the “level of education” variable, the majority of subjects in the study were at secondary school level (60%), while those at university level accounted for only 40%.

Contrary to the study by Malumbu L, in her study of medical care for women in prison, the case of the Kisangani central prison, reported that the majority of inmates had lower or primary education, i.e. 33.3% had secondary education and 30.3% were illiterate. Those with secondary education accounted for 27.3% of cases [18].

In the Democratic Republic of Congo, despite the absence of a job market, the Congolese have become aware of the need to educate their children, based on Unicef's awareness-raising slogan: “All children, girls and boys at school”. As a result, we have a considerable number of high-school students, most of whom are limited to the secondary level for a variety of reasons: early pregnancy, lack of resources, voluntary drop-out, etc.

4.4. Marital status

This study presents the results according to which 14 study subjects, i.e. 70% were married versus 30% single.

These results clearly show that motherhood is an adventure for married people, although some fathers abandon their sickle-cell children. In principle, you can only give birth when you live as a couple. For this reason, more than half of our study population were married.

4.5. Socio-economic level

The study data indicate that 55% of the subjects in the study had a low socio-economic standard of living, while those of socio-economic means represented 35% of cases. On the other hand, two subjects (10%) were of high socio-economic status.

The African Society of Pediatrics, in its Guide to the management of sickle-cell disease in Africa, has shown that sickle-cell disease mainly affects impoverished populations in countries with limited health budgets [19].

In our opinion, many of the advances made in the management of sickle-cell disease are not yet available in the areas where it is most prevalent, but are available to patients in rich countries or to rich patients in poor countries. These advances do exist. We need to make it accessible to as many people as possible.

4.6. Circumstances of discovery of sickle cell disease in children

In Kisangani, episodes of blood transfusion in children and sick children are the circumstances in which sickle cell disease is discovered in 40% and 30% of cases respectively.

In the Democratic Republic of Congo, people have no culture of medical analysis. They wait for the onset and severity of the disease before going to hospital for treatment. Over 60% of the population self-medicate. As a result, chronic illnesses such as sickle-cell anemia, cancer, HIV/AIDS, etc., in both children and adults, are discovered late.

4.7. Number of vaso-occlusive attacks per year

The data from this research indicated that 65% of subjects report several vaso-occlusive attacks per year, while those who have 1 to 4 attacks represent 10% of cases.

A study of sickle cell disease in children in Beni and Butembo, Democratic Republic of Congo, reported that 197 (1.5%) of the patients studied were sickle cell patients. There were 163 cases of severe vaso-occlusive crises (82.7% of sickle cell patients), including 26 not included in this study. A total of 137 cases were included. The mean number of patients hospitalized per year was 15.7 ± 16.5, with extremes of 22 and 30 [19].

In our opinion, the onset of vaso-occlusive crises in sickle-cell patients depends on several factors: hypoxia, cold, fever, dehydration, stress, intake of stimulants, infections, etc. Failure to take these factors into account can lead to vaso-occlusive crises as many times a year.

4.8. Causes of vaso-occlusive attacks

This study shows that infections (malaria, diarrhea and typhoid fever) and climate (cold and heat) are the main causes of vaso-occlusive attacks for 50% and 35% of the respective subjects.

According to HAS, it is recommended to explain to parents the factors favoring painful vaso-occlusive attacks:

- hypoxia: excessive and unusual exertion, altitude (from around 1,500 m), tight clothing, etc. ;
- cooling: cold-water baths, etc. ;
- fever;
- dehydration: vomiting, diarrhoea, etc. ;
- stress;
- intake of stimulants, alcohol, tobacco or illicit drugs (more so in than in children) [25].

The vaso-occlusive crisis is the most frequent; it is provoked by the obstruction of microcirculations by sickle cell red blood cells, responsible for an acute hyperalgesic ischemic lesion of the irrigated organ (mono- or multifocal osteoarticular crises, abdominal crises, thoracic syndrome, etc.) and the resulting pain.

4.9. Elements of care for children with sickle cell disease

Pain control, transfusion in case of anemia, good nutrition and infection control are the main components of sickle cell case management, according to 100%, 85%, 75% and 70% of cases.

According to Keza G.K, Diallo D et al (2023), medical management of sickle cell disease consists of monitoring anemia, preventing painful attacks and infections, and detecting complications early to treat them as quickly as possible. Patients are always provided with a prescription for pain medication [20].

Therapeutic management of sickle cell disease is multidisciplinary, including internists, emergency physicians and resuscitators. It includes symptomatic treatment of vaso-occlusive crises, transfusion, or even exchange transfusion, and organic supplements depending on the severity of the crisis. Patients with severe complications are admitted to intensive care [21].

According to Brillant Damus (2022), the treatment of a simple vaso-occlusive crisis comprises 2 components: analgesic treatment and treatment of factors favoring falciformation. The vaso-occlusive crisis is the cause of very intense bone pain at a level superior to that of a bone fracture. The therapeutic response must therefore be at the same level, and in all cases requires major analgesics such as morphine [22].

According to the African Society of Pediatrics (2018), there are only palliative treatments in sickle cell disease. The essence of treatment consists in managing the symptoms resulting from crises. The sickle cell patient needs rest, good oxygen therapy in the case of thoracic syndromes, good hydration in hot weather, and analgesics in the case of pain.

Emergency transfusion remains the only treatment for profound anemia. Folic acid is given for life to facilitate cell renewal [12].

Any sickle-cell complication should a priori be treated in hospital, because of the unpredictable risk of aggravation or association with other conditions that may have been masked at the outset: any bone pain crisis that is abnormal in terms of duration, intensity, resistance to usual initial treatments, fragility of the terrain in which it occurs, or the impossibility of providing adequate analgesic treatment and hydration on an outpatient basis, should be referred to hospital. Furthermore, all extra-osseous complications are systematic indications for hospitalization, with hyperthermia leading the way, as well as thoracoabdominal pain and vaso-occlusive syndromes, and severe anemia. The efficacy of curative management of acute complications of sickle cell disease depends on the quality of initial management of vital distress and proper diagnostic evaluation of patients [24].

Sickle cell disease remains incurable for most sufferers. However, treatments are prescribed on an individual basis by the specialist physician and are indicated according to each patient's risks. It's important to talk to your specialist to understand what treatments are available and their health benefits. The only curative treatment currently available for sickle cell disease is a bone marrow allograft. This involves destroying the patient's own bone marrow stem cells and replacing them, via transfusion, with those of a compatible donor, such as a brother or sister. Bone marrow is the organ that produces blood cells and hemoglobin. Replacing a sickle cell patient's bone marrow with that of a healthy donor can cure the disease. So, to avoid immune complications, the donor is in most cases a family member [23].

Hydroxyurea, also known as hydroxycarbamide and marketed under the brand names Hydrea and Siklos, is a drug used to prevent and reduce the occurrence of vaso-occlusive crises. This treatment increases the amount of fetal hemoglobin produced by the body. This hemoglobin becomes available in the red blood cells of people with sickle cell disease in sufficient quantities to counterbalance the effects of hemoglobin S, which causes red blood cells to become rigid and crescent-shaped [26].

We believe that optimal care, as provided in reference centers, usually in capital cities, transforms patients' lives. Many reach adulthood, go to school, have a job and start a family. In our opinion, to achieve this, we need to: break the cloak of silence by informing sufferers, families, the public and political decision-makers about the reality of this disease; screen from birth or in the first few weeks of life to considerably improve the quality of life of affected children and avoid certain complications through simple actions; take proper care of sufferers. In our opinion, hydroxyurea is one of the main treatments for improving the course of the disease in sickle-cell patients. Nevertheless, in the Democratic Republic of Congo, its availability could be improved, particularly in a remote town, and its price remains too high for the local population to afford.

4.10. Difficulties experienced by parents in caring for their sickle-cell-affected children

Insufficient financial means and abandonment of children by their fathers are the main difficulties experienced by parents in caring for their sickle cell children, according to 100% and 80% of the respective subjects.

As most parents of sickle-cell children have low incomes, financial limitations are a major problem in caring for their children. The purchase of certain products (hydroxyurea, etc.), obtaining transfusion bags, paying the costs of several hospitalizations, and ensuring a diet in accordance with dieticians all constitute a sea to drink for these unfortunate families. As a result, these children rarely celebrate their fifth birthday.

4.11. Proposed solutions to improve sickle cell disease management

The government should take charge of sickle-cell-affected children by setting up national sickle-cell treatment centers throughout the Republic (100% of cases), and separate A/S and A/S or A/S and S/S couples (65% of cases). However, 30% of subjects reported sterilization of couples to prevent the procreation of sickle cell children.

After an initial “Initiative drépanocytose Afrique” meeting in March 2019, a second meeting was organized on June 15, 2022 in conjunction with the 4th World Congress on Sickle Cell Disease, the Pierre Fabre Foundation (2019) resulted in the publication of a Joint Declaration for the consideration of sickle cell disease by the international community signed by representatives from 12 African countries and Haiti. To achieve lasting improvements in the care of sickle cell patients, the Pierre Fabre Foundation has launched “E-drépanocytose”, a free training platform for healthcare professionals in French-speaking Africa. The first training platform dedicated to the disease, it is in line with the prospect of an increase in the sickle-cell population linked to demographic growth in Africa, and a growing need for staff trained in its management [25].

Effective ways of improving the health of sickle-cell-affected children include: 1. standard treatments: antibiotics, vaccines, folates; 2. treatment of vaso-occlusive crises: analgesics and oxygen therapy; 3. prevention of factors triggering crises (cold, altitude, infections, dehydration); 4. Take folate-rich medication for anemia; 5. Vaccination to prevent pneumococcal and meningococcal infections; 6. Blood transfusion in cases of severe anemia or infection; 7. Blood transfusion to reduce the proportion of hemoglobin S [27].

We believe that in most countries where sickle cell disease is a major public health problem, there are no national programs to combat it. The basic structures needed to care for patients are generally lacking. Systematic screening for sickle cell disease is a simple blood test, yet this is not common practice. In developing countries, the disease is usually diagnosed only when a serious complication occurs. As a result, over 50% of children with the most severe form of the disease die before the age of five, most often from infection or severe anemia. In our opinion, early detection of the disease is essential to enable couples to benefit from good information, education about the disease and appropriate care.

5. Conclusion

The problem of parents caring for sickle cell children at the center Gracia Foundation'' treatment in the city of Kisangani, in the Democratic Republic of the Congo from 15 /10 to 15 /11 2024".

After tabulation and analysis of the data, the following result was observed: insufficient financial means and abandonment of children by their fathers are the main difficulties experienced by parents in caring for their sickle-cell-affected children, according to 100% and 80% of the respective subjects.

Sickle cell disease is a reality in the Democratic Republic of Congo, particularly in Tshopo Province, where parents say that "it's hard to have a sickle cell child". Difficult living conditions expose sickle-cell sufferers to a number of factors that contribute to crises, including difficult access to drinking water, a hot and humid climate, public insalubrity, non-adherence to treatment due to denial of the disease, and insufficient financial resources on the part of parents.

It is important to create culturally and locally relevant interventions to improve access to healthcare, reduce social and cultural barriers, promote a better understanding of the disease and integrate a routine follow-up model for sickle cell patients in other secondary structures, which implies its integration into the health system of the Democratic Republic of Congo.

This also implies that the Congolese state must pay its civil servants well enough to enable them to take proper care of their sick children. Sickle-cell disease affects the general well-being and quality of life of individuals, having a negative impact on social and professional life, school work, and the physical and mental health of patients.

However, the Government of the Republic must put in place programs focused on advocacy, prevention, counseling, screening, early treatment and research, so that children are screened at birth and treated if found to have sickle cell disease. In fact, this simple diagnosis followed by treatment considerably reduces symptoms. It would also be important for adults without sickle cell disease to be screened to find out if they are healthy carriers of the disease, so that a couple will know if there is a risk of having a sick child.

Compliance with ethical standards

Disclosure of conflict of interest

No conflicts of interest have been noted.

Statement of informed consent

Informed but verbal consent was obtained from all individual participants included in the study.

Authors' contributions

This work was carried out in collaboration among all authors read and approved the final manuscript.

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