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Factors the onset of SS anemia and future prospects in the management of sickle cell patients' management of sickle-cell anemia patients

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Abstract

Introduction: Sickle cell anemia is a frequent and serious genetic disease affecting immigrant populations of African and West Indian origin. It is caused by an abnormality in hemoglobin, the protein inside red blood cells that transports oxygen throughout the body, resulting in the deformation of red blood cells, which lose their rounded shape and become banana-shaped.

The objectives of this study are to Identify the various factors favoring sickle cell crisis; Propose the necessary means to prevent sickle cell crisis; Raise awareness of the methods and techniques for preventing sickle cell crisis.

Methodology: This is a descriptive, cross-sectional, quantitative study involving 18 parents of sickle-cell children followed up at the ''Gracia fondation'' center in the city of Kisangani from 15/10 to 15/11 2024. Data were collected using an interview technique based on a pre-established questionnaire.

Results: After analysis, we arrived at the following results: 1. Infections (malaria, arthritis, etc.), climate change and dust are factors favoring sickle cell crises according to 77.7%, 61.1% and 55.5% of the respective subjects; 2. Early treatment of infections, chemoprophylaxis with folic acid and ensuring a good protein diet were cited as preventive measures for sickle cell crises according to 72. 2%, 66.6% and 50% of the respective subjects; 3. Avoiding the marriage of AS/AS couples through premarital examinations (72.2%), requesting the avoidance of maternity in the event of the inadvertent marriage of AS/AS couples (55.5%) and effecting the divorce of AS/AS, SS/SS couples (27.8%) are the methods used to prevent the transmission of sickle cell disease.

Keywords: Factor; Favoring; Crisis; SS Anemia; Sickle Cell

1. Introduction

Following the discovery of hemoglobin S (HBS) by Pauling in 1949, a coherent pathophysiological pattern emerged in the 60s and 70s, detailed at the molecular level. This pattern accounts for hemoxy-hbs anemia, which forms large fibers inside the red blood cell (RBC) that deform and weaken it, and the onset of this crisis is often linked to certain environmental, social and medical factors.

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Sickle cell disease, also known as sickle cell anemia, is a frequent and serious genetic disease, affecting mainly immigrant populations of sub-Saharan African and West Indian origin. It is caused by an abnormality in hemoglobin, the protein inside red blood cells that transports oxygen from the lungs to all the body's organs. This hemoglobin anomaly results in the deformation of red blood cells, which lose their rounded shape (like a disk flattened in the middle) and take on the shape of a banana (a peculiarity which gives the disease its name). This is known as falciformation [1].

According to Khellaf, to suffer from sickle cell disease, both parents must have at least one copy of the S mutation, and pass it on to their children. They cannot present symptoms if they are both A/S. In this case, their children will have a one in 4 (25%) chance of being S/S and, therefore, of suffering from sickle cell disease [2].

Sickle cell disease is a genetic disorder affecting 15,000 patients in France (20,000 by 2020). It has become the leading genetic disease ahead of cystic fibrosis, with more than 50% of sickle cell patients living in the Île-de-France region, and represents a real public health problem [3].

Elion (2021), constant that, in the case where one of the two parents suffers from this disease (he/she is therefore S/S), the risk of transmission is 50% (one chance in two), if one of the parents is A/S and the other is A/A, the children will be 50% AA and 50% A/S, carriers of the mutated gene but without symptoms [4].

Recent epidemiological data show that in the neonatal period, 2% of newborns are homozygous for the disease, and around 40,000 sickle cell births are estimated each year, while the adult population sharing the trait amounts to 25%, with the homozygous form affecting around 2% of individuals [5].

In families where cases of sickle cell disease have been diagnosed, future parents can benefit from genetic counseling and undergo premarital examinations before concluding their marriage or even before conceiving their first child (with analysis of their genes to find out whether they are carriers of the S mutation) [6].

Physical factors that favor the expression of the disease are: dehydration, stress and infectious syndromes cause an increase in the concentration of HbS in red blood cells, while hypoxemia, acidosis and hyperthermia favor deoxygenation of red blood cells. Sickle cell disease often causes a major economic imbalance in 79% of families whose member suffers repeated crises and requires appropriate care. Socio-demographic, economic, cultural, medical-health and environmental factors are all thought to be responsible for the onset of sickle-cell crises [7].

In the course of deoxygenation following certain respiratory dysfunctions, the HBS molecule undergoes a conformational change. The replacement of hydrophilic β6 glutamic acid (a water-soluble substance) by hydrophobic valine causes the latter to establish hydrophobic bonds (a substance or portion of a molecule that does not dissolve in water) with other hydrophobic residues on the β chain of another desoxy-hbs molecule, a polymer is formed and elongated into helical fibers that bundle together, stiffen and cause the characteristic sickle cell shape of classic sickle-shaped red blood cells on the one hand, and on the other, at low oxygen pressure, desoxy HBS polymerizes and organizes into large polymer fibers that deform, stiffen and weaken the red blood cell. This process represents the mechanistic basis of hemolytic anemia and vaso-occlusion [8].

In obstetrics, the occurrence of pregnancy in a sickle cell patient, whatever the genotype (SS, SC, SD, S6 Thalassemia), is a very high-risk situation, as it is marked by high maternal morbidity and mortality in the perinatal period. This association has reciprocal influences: (in sickle-cell anemia, pregnancy aggravates chronic anemia, the high frequency of infections, and the vaso-occlusive crisis at the end of pregnancy, during labor and on the first day of post-partum. In Revenge, sickle cell disease influences delayed menarche, heavy menstruation and/or metrorrhagia which can aggravate anemia, and other obstetric complications including preeclampsia and retroplacental hematoma can occur [9].

In its 2006 report, the WHO estimated that 500 million people carry the sickle cell trait, and that around 50 million people live with the disease. Every year, 300,000 children are born with the disease, 2/3 of them in sub-Saharan Africa. Sickle cell disease is characterized by painful attacks and hematological crises, leading to a high transfusion risk and a high susceptibility to infections. This explains the high morbidity and mortality recorded in sickle-cell patients, and the fact that 50-80% of children born on the African continent will not reach the age of 5 [10].

While vaso-occlusive crisis (VOC) and acute chest syndrome are the most frequent manifestations, the therapeutic bases and other complications such as priapism and stroke need to be known (Brillant DAMUS, 2022) [11].

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-affected 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. [12]. According to Shongo et al (2020), in his study at Clinique NGALIEMA, infections, dehydration, hypoxia, abrupt change of infections (especially pneumopathies), chest pain and low temperature can trigger sickle cell crisis syndromes in 37% of cases [13].

Sickle cell disease remains a major public health problem in the Democratic Republic of Congo, the country with the highest sickle cell burden in the world. In particular, the fact that the DRC is the second country in Africa with the highest number of sickle-cell sufferers, as well as being the first country in the world with the highest number of sickle-cell sufferers with Bantu haplotypes, makes this the most severe clinical form of the disease. The main acute sickle cell complications are vaso-occlusive bone crisis, acute thoracic syndrome, stroke, acute priapism and acute anemia [14].

It is a disease of psychological suffering and isolation. The burden of sickle cell disease weighs on the shoulders of the whole family group, who share the ordeal of the chronic disease and its financial consequences. Patients feel excluded by their handicap and guilty for the imbalances and tensions induced by their misfortune.

With this in mind, the following fundamental questions were posed:

- What factors favor sickle cell crisis?
- What can be done to prevent sickle cell crisis?
- How can the transmission of this genetic disease within the population be prevented?

Research objectives

General objective

The general objective of this research project is to identify the risk factors for preventing sickle cell crises by improving the living conditions of sickle cell patients.

Specific objectives

- Identify the various factors favoring sickle cell crises;
- Suggest ways of preventing sickle-cell crises;
- Raise public awareness of methods and techniques for preventing sickle-cell crises.

1.1. Research goal

The ultimate aim of this research is to raise parents' awareness of the factors that contribute to sickle cell crises, and to promote the well-being of sickle cell sufferers. The aim is to secure the health of the whole community, a source of development.

1.2. Interest of the study

Our work is of threefold interest:

- It will help us to deepen our knowledge of the various factors that contribute to sickle-cell crises;
- This research is a data bank for future researchers who want to undertake a study of aspects not covered in our research;
- It is also a means of raising community awareness to effectively combat the factors contributing to the onset of sickle-cell crisis, and also to prevent the onset of this genetic disease within the community.

1.3. Type of study

This is a descriptive, cross-sectional, quantitative study of factors favoring the occurrence of SS anemia crisis in sickle cell patients at the ''Gracia fondation'' treatment center in the city of Kisangani, Democratic Republic of Congo from 15 /10 to 15 /11 2024.

1.4. Study population

The population of this study is composed solely of parents of sickle cell children attending the ''Gracia fondation'' Sickle Cell Treatment Center in the city of Kisangani. As they rotate, it was difficult to bring them together, so we drew a representative sample to facilitate the study.

Given the importance of this study, our sample included 18 parents who had shown good faith in responding to our questionnaire.

1.5. 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 our study population, we opted for the interrogatory method, whose interview technique enabled us to collect the data. We used a questionnaire which we developed and administered or submitted to the respondents, in the form of a structured interview.

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.

We therefore randomly administered data to 18 parents, making up the size of our study population.

1.6. 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.

2. Results

2.1. Socio-demographic data

2.1.1. Gender

Table 1 Gender distribution of study subjects

Gender	f	%
Female	12	66.7
Male	6	33.3
Total	18	100

Analysis of Table I shows that 66.7% of study subjects were female, compared with 33.3% male.

2.1.2. Age

Table 2 Age distribution of study subjects

Age range in years	f	%
20 - 29	5	27.8
30 - 39	6	33.3
40 - 49	4	22.2
50 and over	3	16.7
Total	18	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 22.2% of cases.

2.1.3. Level of education

Table 3 Distribution of respondents by level of education.

Level of education	**f**	**%**
University	10	55.6
Secondary	8	44.4
Total	18	100

For the "level of education" variable, analysis of Table III shows that the majority of study subjects were university graduates, accounting for 55.6% of cases, while secondary school graduates represented only 44.4% of subjects.

2.1.4. Marital status

Table 4 Distribution of respondents by marital status.

Marital status	***f***	***%***
Married	12	66.7
Single	6	33.3
Total	18	100

This study revealed that 12 subjects (66.7%) were married, compared with 33.3% who were single.

2.1.5. Socio-economic level

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

Socio-economic level	**f**	**%**
Medium	10	55.6
Low	7	38.8
High	1	5.6
Total	18	100

The data in Table V show that 55.6% of the subjects in the study had an average socio-economic standard of living, while those with low socio-economic status accounted for 38.8% of cases. On the other hand, only one subject (5.6%) had a high socio-economic standard of living.

2.2. Variables studied

2.2.1. Knowledge of sickle cell disease

Table 6 Distribution of subjects according to their knowledge of sickle cell disease

Knowledge of sickle cell disease	**f**	**%**
Yes	13	72.2
No	5	27.8
Total	18	100

The data in Table VI show that 72.2% of subjects had some knowledge of sickle cell disease, compared with 27.8% of subjects with insufficient knowledge of sickle cell disease.

2.2.2. Causes of sickle cell disease

Table 7 Distribution of study subjects by cause of sickle cell disease

Causes	**f**	**%**
Witchcraft or curse	2	11.1
Curse	2	11.1
Undecided	5	27.8
Union of couples with AS/AS genes	9	50
Total	18	100

The results of this table show that the union of couples with AS/AS genes is the main cause of sickle cell disease according to 50% of the subjects studied, whereas 27.8% of the subjects knew nothing about it.

2.2.3. Factors favoring sickle cell crisis

Table 8 Distribution of respondents by significance of factors favoring sickle cell crisis (N=18)

Factors favoring sickle cell crisis	**f**	**%**
Infections (malaria, arthritis, etc.)	14	77.7
Climate (season)	11	61.1
Dust	10	55.5
Dehydration	8	44.4
Undecided	5	27.8
Family and cultural factors	4	22.2

Infections (malaria, arthritis, etc.), climate change and dust are factors favoring sickle cell crises according to 77.7%, 61.1% and 55.5% of the respective subjects.

2.2.4. Measures to prevent attacks

Table 9 Distribution of subjects according to preventive measures for sickle cell crises N=18

Measures to prevent attacks	**f**	**%**
Early treatment of infections (malaria, arthritis, typhoid fever, etc.)	13	72.2
Folic acid chemoprophylaxis	12	66.6
Good protein diet	9	50
Avoid exposure to risk factors	7	38.8
Undecided	5	27.8

Table IX shows that early treatment of infections, folic acid chemoprophylaxis and a good protein diet were cited as preventive measures against sickle cell crises by 72.2%, 66.6% and 50% of subjects respectively.

2.2.5. Prevention of transmission of sickle cell disease

Table 10 Distribution of respondents according to measures taken to prevent the transmission of sickle cell disease (N =18)

Prevention of transmission of sickle-cell disease f %	**f**	**%**
Prevent the marriage of AS/AS couples through premarital examinations	13	72.2
If AS/AS couples marry inadvertently, avoid maternity	10	55.5
Pray to God with faith to eliminate S genes in couples	6	33.3
Divorce for AS/AS couples	5	27.8

For the variable "prevention of transmission of sickle cell disease", 72.2% of subjects suggested avoiding the marriage of AS/AS couples through premarital examinations, and 10 respondents, i.e. 55.5% of cases, had asked to avoid maternity in the event of the inadvertent marriage of AS/AS couples. However, 5 respondents, i.e. 27.8%, had asked for AS/AS couples to be divorced.

3. Discussion and comments

3.1. Gender

The study indicated that 66.7% of the subjects were female, compared with 33.3% male.

Yenga, in his study conducted in 2024 on the phenomenon of premature sexuality and its corollaries among adolescents in the Tchai district, reported that young adolescent girls were more likely to engage in premature sexuality, with 60% compared with 40% of adolescent boys [15].

This may be explained by the fact that women are more permanent at home than men, more receptive making them especially accessible to answer research questions. They are the ones who welcome home visitors and respond to their concerns.

3.2. Age

This research has shown that the study population is made up more of subjects in the 20-39 age bracket, i.e. 61.1%, while those aged 50 and over represent only 22.2% of cases.

In the study by Moussa Turé (2009) on the attitudes and practices of couples with primary infertility, 74% were aged between 33 and 42 [16].

By comparing these two results, we can predict that motherhood is an adult adventure. In principle, you can only give birth when you've reached adulthood. For this reason, our study population was made up of the parents of children with sickle cell disease.

3.3. Educational level

For the variable "level of education", the study revealed that the majority of subjects in the study were university graduates with 55.6% of cases, while those with secondary education represented only 44.4% of subjects.

In the study by Moussa Turé (2019) on the attitude and practice of a couple with primary infertility, 53.4% were aged between 33 and 42 [16].

For Yenga, 28 young people, or 43.1% were in secondary school followed by 35.4% in elementary school [15].

While Malumbu L, in her study of medical care for women in prison, the case of Kisangani central prison, reported that the majority of inmates had lower or primary education, with 33.3% in secondary school and 30.3% illiterate. Those with secondary education accounted for 27.3% of cases [17].

3.4. Marital status

This study revealed that 12 subjects, or 66.7%, were married, compared with 33.3% who were single.

These results clearly show that motherhood is an adventure for married people. 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.

3.5. Socio-economic level

The research data showed that 55.6% of the subjects in the study had an average socio-economic standard of living, while those with low socio-economic status accounted for 38.8% of cases. On the other hand, only one subject (5.6%) had a high socio-economic standard of living.

In its Guide de prise en charge la drépanocytose en Afrique (Guide to the management of sickle-cell anemia in Africa), the Société Africaine de Pédiatrie (African Pediatric Society) has shown that sickle-cell anemia mainly affects poor populations in countries with limited healthcare budgets. Many of the advances made in its management are not yet available in the areas where it is most common, but are available to patients in rich countries or to rich patients in poor countries. These advances do exist. It must be made accessible to as many people as possible [14].

In the Democratic Republic of Congo, despite the absence of a job market, the Congolese have become aware of the need to get their children into school, based on Unicef's awareness-raising slogan: “All children, girls and boys at school”. As a result, we have a considerable number of university and 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.

3.6. Sickle cell disease knowledge

The data from this research indicate that 72.2% had knowledge of sickle cell disease, compared with 27.8% of subjects with insufficient knowledge of sickle cell disease.

In sickle cell disease, the red blood cells become rigid and sticky, taking on the shape of a C shaped farming tool called a “sickle”. The sickle-shaped red blood cells die off early, causing their lack in the blood to occur regularly [18].

This situation can be explained by the fact that, in the Democratic Republic of Congo, sickle-cell anemia is a present, incurable disease, attracting so much attention that it is known by everyone for its painful vaso-occlusive crises, and impoverishing patients' families.

3.7. Cause of sickle cell disease

The results of this study report that the union of couples with AS/AS genes is the main cause of sickle cell disease according to 50% of the subjects in the study, whereas 27.8% of the subjects knew nothing about it.

Sickle cell disease is due to a single point mutation in the DNA of the gene coding for beta-globin, located on chromosome 11. It confers a modified structure that enables hemoglobin to form chains (polymers) when the concentration of oxygen in the blood is low (hypoxia). These are the genes responsible for hemoglobin production. The usual gene is called A, and the sickle-cell gene is called S. A person is AA if both parents have passed on the A gene [19].

In our view, sickle cell disease is a hereditary blood disorder caused by the production of an abnormal hemoglobin (Hb), called HbS, which deforms the red blood cells, making them fragile and rigid, leading to occlusive seizures, anemia, and so on.

3.8. Factors favoring sickle cell crises

Infections (malaria, arthritis, etc.), climate change and dust are factors favoring sickle cell crises according to 77.7%, 61.1% and 55.5% 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).

It is also advisable to remind them of the need for abundant hydration ("children should keep their urine as clear as possible"). Abundant hydration is essential. It must be unrestricted and continuously encouraged. It is recommended that parents, and then the child, be informed that the child should drink until the urine is "as clear as possible" [20].

The Société Africaine de Pédiatrie has identified several factors likely to affect the polymerization process: the first is any change, however slight, in the intra-erythrocyte concentration of hemoglobin S molecules; the second is the interruption of polymer growth by hemoglobin F (HbF) molecules intercalated in the fiber. All the factors that have an influence on these parameters are likely to be involved in the pathophysiology of the disease. Such is the case with α thalassemia, frequent in the same populations as those at risk for sickle cell disease. [14].

Moreover, the particular susceptibility to infection is partly the result of progressive auto-splenectomy (functional exclusion of the spleen) and a reduction in the phagocytic capacity of polynuclears. Deformed red blood cells are dehydrated, hyper-concentrated and hyper-aggregable. They thus increase blood hyperviscosity and stasis, which in turn aggravate hypoxia and acidosis. The result is a vicious circle. These various vaso-occlusive, hemolytic and infectious phenomena are often interlinked. They are responsible for the clinical manifestations and acute and chronic complications of sickle cell disease [14].

In patients with major sickle cell syndromes, malaria is a factor in the morbidity and lethality of sickle cell disease for at least two reasons: the fever that accompanies it is, in itself, a trigger for vaso-occlusive crises. The onset of pregnancy in a patient with sickle cell disease, whatever her genotype, is a very high-risk situation, marked by high maternal and perinatal morbidity and mortality. This association requires management by a multidisciplinary team in a suitable center [20].

According to Khellaf (2015), cold, altitude, air travel, stress, school or university exams, etc., can affect the heme chain in stressful situations (infections, hypoxemia, acidosis), causing red blood cells to falciform, leading to occlusion of blood capillaries and particularly painful bone infarctions, which explain the vaso-occlusive crises that bring patients to the ER [2].

According to Ndikumana (2020), in obstetrics, pregnancy influences sickle cell crisis by aggravating chronic anemia, the high frequency of vaso-occlusive crises and the high rate of urinary tract infections, pneumococcal infections and surgical site infections in the case of Caesarean sections. On the other hand, sickle cell disease is also associated with delayed menarche up to the age of 18, preeclampsia and retroplacental hematoma [21].

In the practice of anaesthesia in sickle-cell patients, certain anaesthetic products cause respiratory distress which can reduce oxygen levels (hypoxia), resulting in a consequent sickle-cell crisis [22].

In our opinion, there are several factors that favor the expression of the disease, notably: dehydration, which causes an increase in the concentration of HbS in the red blood cells and vaso-construction of the vessels; hypoxemia; acidosis; climent (cold), which favors vaso-construction of the vessels; hyperthermia, which favors deoxygenation of the red blood cells; and infections (in this case malaria). Parents must therefore educate their children to avoid them.

3.9. Seizure prevention measures

The results of this study show that early treatment of infections, chemoprophylaxis with folic acid and a good protein diet were cited as measures to prevent sickle cell crises by 72.2%, 66.6% and 50% of subjects respectively.

According to the Ministry of Public Health and the Fight against AIDS (2020), prevention of sickle cell crises consists in taking abundant beverages and avoiding : - rapid variations in temperature, - taking iced drinks, - violent or sustained exertion, - travelling in non-pressurized aircraft, - staying in confined spaces It is important to get the child into the habit of frequently drinking water and other sweetened beverages. In addition: - Follow the EPI-recommended vaccination schedule, and vaccinate against the germs most frequently responsible for sickle-cell infections: Antipneumococcal, Antimeningo A and Co Antityphoid (typhimVi), 3 doses of hepatitis B vaccine are recommended, - Penicillin V 50mg/kg/d until 5-7 years of age, good personal hygiene and systematic deworming with albendazole or mebendazole every 3 months [22].

Folic acid, or vitamin B9, plays an essential role in cell renewal, particularly of red blood cells. In sickle cell disease, red blood cells are renewed much more rapidly than in non-diseased individuals. Specialists therefore suggest that people with sickle cell disease take folic acid on a regular basis to facilitate this cell renewal [13].

For the African Pediatric Society, preventive measures for vaso-occlusive crises in sickle cell disease include: wearing clothing that does not cut off blood circulation (i.e. loose-fitting), not getting out of breath, avoiding going to altitudes of over 1,500 metres, and not exposing oneself to extreme heat (dehydration triggers crises by increasing blood viscosity) [14].

Regularly prescribed hydroxycarbamide reduces the frequency of painful attacks and hospitalizations, and avoids certain complications. It is an inexpensive raw material. Antibiotic prophylaxis with penicillin V is recommended in children with SS sickle cell disease, to control the risk of infection at each medical consultation or visit, and to reduce progressive non-compliance with treatment. It is also recommended that children with sickle cell disease take oral paracetamol, and that they be vaccinated according to the vaccination schedule against diphtheria, tetanus, poliomyelitis, whooping cough, Haemophilus inflenzae type B infections, rubella, mumps, measles, tuberculosis and hepatitis B [20].

In our opinion, it is important to educate parents of sickle-cell-affected children to be vigilant in avoiding at all costs the factors that trigger attacks (lack of sleep, late bedtime, infection, trauma, taking alcohol, illicit drugs, testosterone or psychotropic drugs, being up to date with vaccinations, decontrolling nutrition, etc.).

3.10. Prevention of transmission of sickle cell disease

For the variable “prevention of sickle-cell transmission”, 72.2% of subjects suggested avoiding the marriage of AS/AS couples through premarital examinations, and 10 respondents, or 55.5% of cases, had asked to avoid maternity in the event of the inadvertent marriage of AS/AS couples. However, 5 respondents, i.e. 27.8%, had asked for AS/AS couples to be divorced.

For Burundi's Ministry of Public Health and the Fight against AIDS, prevention can be achieved by the systematic introduction of hemoglobin electrophoresis as a premarital examination for every couple at risk. It is important to provide genetic counseling for at-risk couples and to advise against marriage between at-risk individuals, i.e. between AS and AS, AS and SS, SS and SS [22].

According to Livi France's Medical Department, it is not possible to prevent the onset of a hereditary disease, but it is possible to limit the onset and severity of complications by several means:

- monitoring and early detection of the disease

- up-to-date vaccinations. In addition to the usual vaccinations, people with sickle cell disease need to be vaccinated against meningococcal ACYW135, influenza and pneumococcal disease, as well as hepatitis A, typhoid and other infectious diseases, depending on the destination;

- Preventive antibiotic treatment in childhood to limit the risk of mortality from serious infection;

- establishment and observance of daily rules for patients: good personal hygiene (brushing teeth, washing hands before each meal) to avoid infections; maintaining good hydration; avoiding chills (bathing in cold water below 25°C, not applying ice packs in the event of a fall); protecting against heat (protecting against the sun); avoiding intense or violent physical activity; avoid reducing oxygen levels in the blood (altitude above 1500m, scuba diving); avoid anything that might block blood circulation; eat a balanced and varied diet; avoid smoking and alcoholic beverages; get enough sleep and reduce stress levels; avoid self-medication, etc. [23].

We believe that sickle-cell anemia is an inherited genetic disease with so-called autosomal recessive transmission: each parent must pass on the mutated gene to the child in order for the disease to develop in the latter. To prevent the transmission of sickle-cell anemia, we advise against marriage between AS and AS, AS and SS, SS and SS couples by means of premarital hemoglobin electrophoresis tests. Consequently, when the fetus has received a single mutated allele, it is a healthy carrier.

4. Conclusion

This study is a descriptive cross-sectional study on "Factors favouring the onset of SS anaemia and future prospects in the management of sickle cell patients at the ''Gracia fondation'' center in the town of Kisangani, Democratic Republic of Congo, from 15/10 to 15/11 2024".

After data processing and analysis, the following main results were observed:

- Infections (malaria, arthritis, etc.), climate change and dust are factors favoring sickle cell crises according to 77.7%, 61.1% and 55.5% of the respective subjects;
- Early treatment of infections, chemoprophylaxis with folic acid and a good protein diet were cited as preventive measures against sickle cell crises by 72.2%, 66.6% and 50% of subjects respectively;
- Preventing the marriage of AS/AS couples through premarital examinations (72.2%), requesting the avoidance of maternity in the event of the inadvertent marriage of AS/AS couples (55.5%) and divorcing AS/AS, SS/SS couples (27.8%) are the ways of preventing the transmission of sickle cell disease.

Sickle-cell disease is a reality in the Democratic Republic of Congo, particularly in the Tshopo Province, in the city of Kisangani, where 80% of sickle-cell-affected children do not celebrate their fifth birthday. What's more, sickle-cell anemia occurs, in most cases, in girls with fewer resources, and caring for these sick children poses serious problems. It is therefore important and necessary to raise awareness of premarital examinations (hemoglobin electrophoresis) among young people wishing to marry, to limit the worst.

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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