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Sickle cell trait, knowledge, attitudes, practices and perceptions regarding sickle cell disease among people living in Yaoundé, Cameroon

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ABSTRACT

Sickle cell disease is the most common autosomal recessive disease in sub-Saharan Africa. Our study aimed to determine the sickle cell trait, to assess knowledge, attitudes, practices and perceptions regarding sickle cell disease of people living in Yaoundé. We carried out a cross-sectional and descriptive study at the Institute of Medical Research and Medicinal Plants studies in Yaoundé. We included a total of 191 participants of mixed sex and ages ranging from 10 to 70 years old, able to complete a questionnaire and having accepted a blood sample for the hemoglobin electrophoresis test. Data was collected using a questionnaire. We used the Chi-square test to assess the relationships between variables with p-value < 0.05 for meaningful relationships. More than half of the participants were women (59.16%), and the representative age group was 20-29 years old (47.12%). The highest level of education was university level (71.73%). Most of the participants had heard of sickle cell disease (93.72%) and the most talked-about information channel was television (41.90%). The proportions in relation to knowledge about sickle cell disease were as follows: in the transmission mode, 71.73% had mentioned that it is an inherited disease; in prevention mode, 88.08% had mentioned the hemoglobin electrophoresis test. For the follow-up of the disease, 85.86% noted the hospital as the location. Compared to marriage with a sickle cell disease patient, 68.59% answered negatively. There is a significant relationship ($p<0.05$) between the level of education and knowledge of sickle cell disease transmission. The results of our study showed that the university population of Yaoundé had a good knowledge of sickle cell disease. Young people are called upon to be tested for sickle cell disease before marriage. Studies on larger samples should be conducted to have a better appreciation of the sickle cell trait in the population of Yaoundé.

Keywords: Sickle cell trait, knowledge, attitudes, practices, sickle cell disease, Center Region, Yaoundé Cameroon.

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INTRODUCTION

Sickle cell disease is a chronic disease, autosomal recessive genetics linked to abnormal hemoglobin (Ebakisse-Badassou, 2010). It is a point mutation of adenine by thymine at the sixth codon of the beta-globin

chain (on chromosome 11); thus causing the substitution of glutamic acid by valine in the sixth position of the beta chain of globin (Baledent and Girot, 2016; Arishi et al., 2021). This mutation is a structural variant of normal adult hemoglobin (Hb AA). The resultant hemoglobin S (Hb SS), which polymerises under low oxygen tension, causes rigid, sickled red blood cells (Rees et al., 2010). The genetic defect of sickle cell disease is spread according to Mendel's laws (Baledent and Girot, 2016). Checking for abnormal hemoglobin and confirmatory diagnosis are based on biological techniques (Wajcman and Moradkhani, 2011). According to World Health Organization (WHO), more than 120 million people carry the sickle cell trait on all continents of the world and about 500,000 infants are born homozygous SS each year. It is the most common genetic pathology in the world (WHO, 2010). It is very common in the black race and it is a public health problem (Mpiana et al., 2016). In Central and West Africa, 20 to 40% of people are carriers of the sickle cell trait. About 300000 children are born there with sickle cell disease each year. This disease is responsible for around 5% of deaths of children under five on the African continent (Ebakisse-Badassou, 2010). In Cameroon, the prevalence of sickle cell trait is 22.3% (Menick, 2014). The prevalence of SS homozygosity varies from 1.7 to 9% depending on the region. The incidence of sickle cell disease in Yaoundé is estimated at 7.2% (Mpiana et al., 2016). A study carried out at the Essos Hospital Center in Yaoundé on 5,856 newborns screened at birth, revealed an AS trait prevalence of 13.2% and that of the homozygous form SS of 0.1% (Motaze, 2013). Early diagnosis, from birth or in the first weeks of life, can significantly improve the quality of life of affected children and avoid certain complications through preventive measures. Screening for carriers of the trait is an important step in reducing the incidence of the disease. Each informed adult and couple can thus adapt their life plans and avoid having sickle cell children. Our study aims to evaluate the level of knowledge, attitudes and practices of the population of Yaoundé with regard to sickle cell disease, and then to determine the frequency of the AS genotype in this population.

MATERIALS AND METHODS

Type and location of the study

We conducted a cross-sectional and descriptive study from 02 to 31 March 2020 at the Ministry of Scientific Research and Innovation, more precisely at the Human Biology Laboratory of the Institute of Medical Research and Medicinal Plants Studies (IMPM) in Yaoundé. It took place during the campaign days organized by the Institute on the occasion of International Women's Day.

Study population

We recruited a total of 191 patients of both sexes. Our sampling was calculated according to the prevalence of sickle cell disease (9%) reported by Menick in a study carried out in Cameroon (Menick, 2014) using the Lorentz formula. Hence our minimum sample size required for this study was 125. We included in the study any patient who came into the human biology laboratory of IMPM during the campaign days, aged 10 to 70, who had a correctly completed questionnaire, and who have agreed to have venous blood taken at the laboratory for hemoglobin electrophoresis testing. Any patient who did not give informed consent and who has not donated enough blood or whose venous sampling was not successful was excluded from the study.

Ethical consideration

Informed consent was obtained in advance from adult participants and minors' consent was obtained from their parents. They were reassured that their results would be anonymous and confidential. Ethical clearance was obtained from the IMPM Ethics Committee and administrative authorization has been obtained from the director of the institute (IMPM) to carry out the study. Ultimately the study was submitted to the National Research Ethics committee for human health (CNERSH) to obtain their approval and ethical clearance was issued under the reference N° 2020/01/752/CE/CNERSH/SP.

Data collection and blood sampling

The data was collected in a questionnaire drafted by the Institute's researchers. For the participants who did not understand the English language, the questionnaire was explained in the French language. This questionnaire was divided into socio-demographic data (age, gender, marital status, etc.), Knowledge about sickle cell disease (Have you ever heard of sickle cell disease, by which information channel, what are the clinical signs of sickle cell disease), attitudes and practices (Do you know your partner's status? do you know your own status, can you marry a sickle cell patient?...), of patients towards sickle cell disease. Then we collected 5 ml of whole blood in tubes containing EDTA (Ethylene diamine tetraacetic acid as an anticoagulant) with sterile single-use equipment and we performed the hemoglobin electrophoresis test on cellulose acetate paper in a SHANDON (2013) electrophoresis tank with tris glycine buffer. We had blood sample controls for hemoglobin AA (normal hemoglobin), hemoglobin AS (hemoglobin of sickle cell trait carriers) and hemoglobin SS (abnormal hemoglobin, hemoglobin of sickle cell disease).

Statistical analysis

Data were entered into excel 2013 and analyzed with Epi Info version 7.2 software. The Chi-2 test and Fisher's exact test for the number below 5 were used to test associations between variables and $p < 0.05$ was retained for significant relationships.

RESULTS

In our study, we had a total of 191 participants who completed the questionnaire correctly. The most represented gender was female (n = 113; 59.16%), the most represented age group was 20 to 29 years (n = 90; 47.12%). More than half were single (n = 139; 72.77%)

and had a university degree (n = 137; 71.73%). The West region was the most represented region of origin (n = 68; 35.60%). And for the number of children n = 111 (58.12%) were without children (Table 1).

Among the 191 participants in our study (n = 179 or 93.72%) who had already heard of sickle cell disease, the most represented information channel was television (n = 75; 41.90%) followed by the school (n = 64; 35.75%) (Table 2). On the question of whether only one parent can transmit the disease to their child n = 73 or 38.22% had answered positively. On disease prevention, n = 151 or 79.06% also responded positively, mentioning hemoglobin electrophoresis as a prevention method (n=133 or 88.08%). In addition, n = 138 or 72.25% had answered positively to the question of whether both parents must be carried to transmit the disease (Table 3).

Table 1. Socio-demographic characteristics.

Variables	N	Percentage (%)
Age (years)	**N=191**	
10-19	11	5.76
20-29	90	47.12
30-39	55	28.80
40-49	21	10.99
50-59	10	5.24
60+	4	2.09
Sex	**N=191**	
Female	113	59.16
Male	78	40.84
Marital status	**N=191**	
Single	139	72.77
Married	52	27.23
Region of origin	**N=191**	
Adamaoua	04	2.09
Centre	46	24.08
Far North	14	7.3
East	04	2.09
Littoral	22	11.52
North West	07	3.66
North	07	3.66
West	68	35.60
South	11	5.76
South-West	08	4.19
Level of education	**N=191**	
Primary school	06	3.14
Secondary school	48	25.13
Higher education	137	71.73
Number of children	**N=191**	
0	111	58.12
1	34	17.80
2	16	8.38
3	09	4.71
4	11	5.76

Table 1. Continues.

5	05	2.62
6	04	2.09
7	01	0.52
8	00	00

Table 2. Sickle cell disease; source of information.

Sickle cell disease	N=Number	Percentage (%)
Have you ever heard of sickle cell disease?	**N=191**	
Yes	179	93.72
No	12	6.28
If yes, through which channel?	**N=179**	
Television	75	41.90
Radio	04	2.23
School	64	35.75
Hospital	16	8.94
Friends and Family	17	9.50
Others	03	1.68

Table 3. Sickle cell disease; transmission, prevention.

Sickle cell disease	N=191	**Percentage (%)**
Hereditary	137	71.73
No-hereditary	19	9.95
I don't know	35	18.32
Just one parent affected can transmit the disease?	N=191	
Yes	73	38.22
No	60	31.41
I don't know	58	30.37
Both parents must be affected to transmit the disease	N=191	
Yes	138	72.25
No	11	5.76
I don't Know	42	21.99
Can this disease be prevented?	N=191	
Yes	151	79.06
No	08	4.19
I don't know	32	16.75
If yes, how?	N=151	
Doing electrophoresis	133	88.08
Medical doctor's advice	15	9.93
I don't know	03	1.99

The most frequently mentioned clinical signs were pain (n = 90; 47.12%) and anemia (n = 55; 28.80%). N = 153 or 80.10% mentioned blood as a means of screening. For the hemoglobin electrophoresis test, n = 134 (70.16%) had stated that they were aware of this test and n = 124 or 92.54% had mentioned that it was used for the diagnosis of sickle cell disease. Unfortunately, n = 144 or 75.39% said they had never done the electrophoresis test before. For those who had already been tested (n = 47 or 24.61) the different genotypes were as follows: AA (n = 34; 80.86%) AS (n = 8; 17.02%) and SS (n = 1 or 2.13%) and the rest (n = 4 or 8.51%) no longer knew the result. For the follow-up of a sickle cell patient, n=164 or 85.86% had mentioned the hospital (Table 4).

Table 4. Clinical signs, diagnosis and practices for sickle cell disease.

Variables	Number	Percentage (%)
What are the clinical signs?	**N=191**	
Pain	90	47.12
Anemia	55	28.80
Yellow eyes	08	4.19
Infections	03	1.57
Necrosis femoral head (Lameness)	01	0.52
Splenomegaly	03	1.57
I don't know	31	16.23
How is sickle cell disease diagnosed?	**N=191**	
Blood	153	80.10
Urine	01	0.52
I don't know	37	19.38
Did you know about the electrophoresis test?	**N=191**	
Yes	134	70.16
No	57	29.84
If yes what is the purpose?	**N=134**	
Screening sickle cell disease	124	92.54
Others diseases	05	3.73
I don't know	05	3.73
Have you ever done a hemoglobin electrophoresis test?	**N=191**	
Yes	47	24.61
No	144	75.39
If yes what is the result	**N=47**	
Hb AA	34	72.34
Hb AS	08	17.02
Hb SS	00	00
I don't know	05	10.64
If you have sickle cell disease where do you go for treatment?	**N=191**	
Hospital	164	85.87
Tradipratician	00	00
Church	01	0.52
I don't know	26	13.61

Out of 191 participants, 159 did not know their partner's status (83.25%); then n = 134 or 70.16% had answered that their choice could be influenced if they knew their partner's status. Regarding the marriage to a sickle cell patient, n = 131 or 68.59% answered negatively. The genotypic proportions of the study results are as follows: AA 96.32%; AS 3.68%; SS 0% (Table 5).

There was a statistically significant relationship ($p < 0.05$) between the level of education and knowledge of sickle cell disease transmission. Academics were statistically more knowledgeable about the modes of transmission of sickle cell disease than primary and secondary school patients (Table 6).

There was a statistically significant relationship ($p < 0.05$) between the level of education and knowledge of clinical signs of sickle cell disease. Academics are more knowledgeable about the clinical signs of sickle cell disease than secondary and primary level patients (Table 7).

There was a statistically significant relationship ($p < 0.05$) between the level of education and knowledge of how to prevent sickle cell disease. Academics are better informed about how to prevent sickle cell disease (Table 8).

Table 5. Perceptions and attitudes towards sickle cell disease.

Variables	Number	Percentage (%)
Do you know the status of your partner?	**N=191**	
Yes	32	16.75
No	159	83.25
Could the sickle cell status influence your choice of sexual partner?	**N=191**	
Yes	134	70.16
No	32	16.75
I don't know	25	13.09
Can you marry a sickle cell patient?	**N=191**	
Yes	29	15.18
No	131	68.59
I don't know	31	16.23
Test result	**N=191**	
Hb AA	184	96.32
Hb AS	07	3.68
Hb SS	00	00

Table 6. Relationship between level of education and knowledge of sickle cell disease transmission.

Level of education	Hereditary	No hereditary	I don't know	Total	P-value
Primary school	1	0	5	6	X^2=35.54 ddl=4 P<0.05
Secondary school	24	8	16	48	
Higher education	112	11	14	137	
Total	137	19	35	191	

Table 7. Relationship between level of education and knowledge of clinical signs of sickle cell disease.

Level of education	Anemia	Lameness	Pain	Infections	bloated belly	Yelloweyes	I don't know	Total	P-value
Primary school	0	0	0	1	0	1	4	6	$X^{2=}$39
Secondary school	9	1	22	0	2	2	12	48	ddl=12
Higher education	46	0	68	2	1	1	15	137	P<0.05
Total	55	1	90	3	3	3	31	191	

Table 8. Relationship between level of education and prevention of sickle cell disease.

Level of education	Doing electrophoresis test	Medical doctor advice	I don't know	Total	P-value
Primary school	0	2	1	3	X^2=32.72
Secondary school	26	6	1	33	ddl=4
Higher education	107	7	1	115	P<0.05
Total	133	15	3	151	

Table 9. Relationship between level of education and knowledge of sickle cell monitoring.

Level of education	Hospital	Church	I don't know	Total	P value
Primary school	5	0	1	6	X^2=1.91
Secondary school	39	0	9	48	ddl=4
Higher education	120	1	16	137	p>0.05
Total	164	1	26	191	

There was no statistically significant relationship (p > 0.05) between education level and knowledge of sickle cell monitoring (Table 9).

DISCUSSION

In sub-Saharan Africa, sickle cell disease is the most common genetic disease. It is the leading cause of chronic hemolytic anemia, posing a real public health problem. The prevalence of hemoglobin 'S' is relatively high. Frequencies greater than 10% have been estimated in Central Africa and West Africa (Piel, 2013).

Socio-demographic data

In our study, the results showed that the most represented age group was 20 to 29 years old or 47.12%, this value could be justified by the fact that the population was predominantly young and were students. Our results corroborate with that of Tusuubira in Uganda who, in their study recruited a predominantly young population (Tusuubira et al., 2018). We had a sex ratio of 0.69 in favour of women. This shows the concern of women for their health, furthermore, it is a laboratory test that is part of the prenatal check-up, and they have free time compared to men who are busier for professional reasons. This is consistent with Guedehoussou in Togo and Ngwengi in Cameroon were in their studies, the sex ratios in favour of women were 0.79 and 0.81 respectively (Guedehoussou et al., 2009; Ngwengi et al., 2020). Our population was predominantly single (72.77%) and as for the level of education, the most represented was university level with a percentage of 71.73%; this can be explained by the fact that most of the participants were students because the Institute for Medical Research and Medicinal Plant Studies is surrounded by a university and several colleges of higher education and these students are mostly single and do not have children 58.12%. Similar results for singles were found in Ghana and Texas with proportions of 55.71% and 70.1% (Orish et al., 2014, Smith and Brownell, 2018).

Knowledge about sickle cell disease

Regarding information on sickle cell disease, 93.72% had already heard of the disease and the main source of information was television (41.90%) followed by the school (35.75%). However, the study by Adewoyin in Nigeria and Tusuubira in Uganda had instead shown that the population was of course informed, but through other channels such as awareness campaigns, hospitals, church, school, family and friends (Adewoyin et al., 2015; Tusuubira et al., 2018); this can be justified by the fact that the majority of households in Yaoundé have a television, in addition, the participants were educated, given their high level of education, they are informed about this disease.

The level of knowledge was assessed on transmission, clinical signs, screening methods and monitoring of the disease. From a general point of view, the participants were quite informed; 71.73% knew that it is an inherited disease from both parents carrying the sickle cell trait. As for the screening method, 72.25% knew that it could be detected by a blood test, namely hemoglobin electrophoresis, which was known by 88.08% of our study population. The clinical signs most frequently mentioned were pain (47.12%) followed by anemia (28.80%) and for the follow-up of the disease, the hospital was the best choice with a percentage of 85.86%. These results showed that the population of Yaoundé is informed about sickle cell disease. In

contrast, in the study by Al-Qattan et al. in Saudi Arabia and Ngwengi et al. in Cameroon, the level of knowledge of the participants was quite low with a percentage of 28.8 and 20.5% respectively (Al-Qattan et al., 2019, Ngwengi et al., 2020)**.** However, there was a statistically significant relationship ($p < 0.05$) between the level of education and knowledge about transmission, prevention and clinical signs of the disease. However, there was no statistical significance between the level of education and knowledge of the disease monitoring ($p > 0.05$).

Attitudes and practices towards sickle cell disease

In our study, 75.39% of the population had not yet done the electrophoresis test. This could be explained by the fact that our population was young, were students, single with a low financial income and 58.12% did not have children.

Although most of the participants had a high level of knowledge about sickle cell disease, this did not affect their perception of the disease, as 83.25% did not know the status of their partner, 70.16% had stated that their status (AA, AS, SS) would influence the choice of a partner, and 68.59% could not marry a sickle cell patient. These perceptions may be due to the fact that sickle cell disease is a fatal disease and the union between two people carrying the sickle cell trait would give a 25% risk of having a sick child in each pregnancy (Baledent and Girot, 2016). Data with the same results and opinions were found in the study by Tusuubira, 90.2% did not know the status of their partner, 60.8% would consider choosing a partner, and 68.3% gave negative answers for marriage to a sickle cell patient (Tusuubira et al., 2018). The results of the screening test gave the following percentages by genotype: AA 96.32%, AS 3.68%, SS 0%. In Cameroon**,** Ngwengi found the following genotypes: AA; 84.0%, AS; 16.0% (Ngwenji et al., 2020) and Boadu in Ghana also found AA; 82.1%, AS; 16.9%, SS; 0.9% (Boadu and Addoah, 2018).

Conclusion

This study showed that the population of the city of Yaoundé with a university degree had a good level of knowledge about sickle cell disease; but, many patients did not know their status. Young people are called upon to be tested for sickle cell disease before marriage in order to avoid unpleasant consequences and parents must test their children for this disease so that they are informed of their status before choosing their future partner. Studies on large populations should be conducted to have a better appreciation of the sickle cell trait and those with sickle cell disease in the population of Yaoundé. The development of public health programs including preventive methods like public education and genetic screening is recommended to control sickle cell disease in these sub-Saharan African countries.

Competing interests

The authors declare that they have no competing interest

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