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Sickle Cell Disease in Southeast Zone of Gabon a Central African Country: A Socio-Economic Study

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Abstract
More than 120 million people worldwide carry the sickle cell trait, and an estimated 500,000 children are born each year with sickle cell disease. This pathology makes no distinction of social class, and its expression is dependent on the presence of the "S" allele in both members of a couple. The aim of this program is to raise awareness about sickle cell disease; to evaluate the level of knowledge and care of sickle cell patients, the level of income of households affected by sickle cell disease and the different needs of sickle cell patients. A series of survey were conducted in the field in each household in the Franceville city council. The data was recorded on WampServer and repatriated on an Excel file, and statistical analysis was performed by R software (version 4.2.1) and its environment RStudio (version 1.1.445). A total of 2777 households responded to our survey. Mothers have a better knowledge of the disease than fathers. Mothers screen more children than fathers and mothers with SS or SC status screen less than other mothers. The number of children screened at birth appears to be associated with income. Households with the lowest incomes screen more of their children, with 87% screening all their children. The population of Franceville was found to be poor overall, with a monthly income of less than 150,000. However, there was no correlation between monthly income and sickle cell disease screening and between monthly income and having a sickle cell child.

Keywords: Sickle Cell Disease, Gabon, SCDOGa, Screening, Socioeconomic Status

1. Introduction

Sickle cell disease is an inherited genetic disease that is transmitted from parent to child. It is caused by a structural abnormality of the beta chain of hemoglobin, resulting in an abnormal form of red blood cells and leading to an obstruction of the blood circulation. People suffering from sickle cell disease remain fragile to bacterial infections.

For several centuries, the fight against many genetic and hereditary diseases throughout the world is still going on, as is the case with sickle cell disease. The history of sickle cell disease is marked by its first observation by James Herrick on a blood smear until the molecular identification of hemoglobin "S" by Venon Ingram in 1956 [1]. Considered the most common genetic disease in the world, sickle cell disease is still largely unknown and often ignored by the population despite the efforts of the last decade to raise awareness about this disease [2].

Indeed, more than 120 million people worldwide carry the sickle cell trait, and an estimated 500,000 children are born each year with sickle cell disease [2]. In Central Africa, one in 30 newborns is sickle cell. Originally widespread in southern countries, it has become common in Western Europe, where in France it mainly concerns individuals from former African colonies [3]. In France, nearly 350 newborns are screened each year [4]. The virulence of sickle cell disease wherever it is found, produces a very strong economic and social impact especially since chronic complications, painful crises and other complications of sickle cell disease are often hidden or ignored and constitute a heavy burden for the family [5].

In many African countries, 10% to 40% of the population carries a sickle cell gene, with an estimated prevalence of sickle cell disease of at least 2% [6, 7]. According to one estimate, approximately 50% to 80% of the 500,000 children born each year in Africa with sickle cell disease die before the age of five [8]. Those who survive have organ damage, which shortens their lifespan [9].

In Gabon, carriers of the sickle cell gene represent 25% (±3) of the population, i.e., one in four Gabonese [10]. About 1.34% of the Gabonese population suffers from sickle cell disease [11], with about 800 homozygous births each year [10]. The prevalence of sickle cell trait carriage is estimated to be 21.1% in individuals over the age of 15 years [12]. According to a study conducted between 2012 and 2017 by national researchers, of the 4,514 samples taken from the Estuary to Woleu-Ntem, the extreme values were recorded in Woleu-Ntem and Haut-Ogooué. This pathology makes no distinction of social class and its expression is dependent on the presence of the "S" allele in both members of a couple [13].

Symptoms of the disease vary in intensity and severity among patients. However, the natural course of sickle cell disease is fraught with many complications, including an increased risk of serious bacterial infections and vaso-occlusive crisis (VOC), which causes pain that is usually sudden and severe [14, 15]; acute chest syndrome (ACS), stroke, priapism, acute abdominal syndrome and worsening anemia. The anemia present in people with sickle cell disease often results in fatigue, weakness and pallor. Eye discoloration, physical deformity, weight loss and even delayed weight gain are also symptoms often associated with this disease [16]. The fragility of their health condition makes them prone to frequent stay in the hospital [7]. All the above symptoms decrease the life quality of not only the patient but also their family, placing a significant emotional, physical and financial burden on them. Indeed, sickle cell disease, beyond the organic damage, causes other sufferings, notably moral distress, isolation, narcissistic wounds, withdrawal and even collapse [16].

In order to improve the follow-up and management of people with sickle cell disease, it is therefore more than essential to promote early detection of cases and to implement a comprehensive management strategy[6]. Sickle cell disease patients are called upon to have regular medical follow-up, a healthy lifestyle, and a balanced diet to reduce the frequency of attacks and minimize the complications associated with this pathology. However, if early treatment of some patients reduces their attacks and increases their life expectancy, for others, several complications appear due to the lack of means and information on the disease and its treatment. Indeed, in Gabon and more specifically in the province of Haut-Ogooué, despite the awareness of the importance of this disease by the public authorities, sickle cell disease remains a poorly known disease among the population. Moreover, prenatal diagnosis is expensive and inaccessible to the majority of couples, neonatal diagnosis is still in its infancy, there is no specifically dedicated service, transfusion treatment is still problematic, vaccines outside of the Expanded Program of Immunization (EPI) and antibiotics are expensive for families with at least one sickle cell patient, most of whom have a very low monthly income. It is therefore appropriate to assess the socio-economic impact of households with at least one sickle cell child in Haut Ogooué province.

This project proposed and implemented by the NGO DrépaZéroCytose Gabon, and it is the first study of its kind in our country, is referenced under the number, SCDOGa/HO1. The objectives of this program are to raise awareness about sickle cell disease; to evaluate the level of knowledge and care of sickle cell patients, the level of income of households affected by sickle cell disease and the different needs of sickle cell patients.

2. Materials and methods

2.1 Consistency of the SCDOGa/HO1 project idea

a) Analysis of the source of the project idea

It was a question of verifying the origin of the source of the project idea before its materialization in project. For this purpose, we defined four sources of project ideas authorized by the management of the NGO SCDOGa, namely: the expression of a need by the population (population targeted by SCDOGa), direct observation of a situation by SCDOGa, recommendations from local authorities (local communities and the state) and technical partners (NGOs, associations, companies). Thus, for the SCDOGa/HO1 project, the authorized sources that were identified corresponded to the first two sources, i.e., the expression by the population targeted by the SCDOGa of a real need to improve their care conditions. This need was identified during the exchanges and direct observations with the populations concerned.

b) Analysis of the logic of the project idea

The verification of the logic of the project idea on the one hand and the consideration of the quality of a project on the other hand were carried out. Indeed, in the case of the SCDOGa/HO1 project, we were able to note that the latter was perfectly in line with the vision and objectives of the NGO SCDOGa because it aimed to support the education and care of sickle cell patients in the city of Franceville. Regarding the approach that consisted of verifying that the project adhered to the quality principles of a good project, we proposed to proceed with an analysis of the qualities of the project. To do this, we used the PERCOEFIDU tool, which considered criteria such as Relevance, Coherence, Efficiency, Feasibility, Impact and Sustainability.

Clarification of the quality of the SCDOGa/HO1 project.

2.2 Study of the strategy for responding to the problem posed by the project idea

The strategy of response to the problem posed by the project idea was defined on four main axes: considering the intelligence and experience of some major actors of the project, considering the local knowledge and know-how and the beneficiaries of the project; the understanding of the purpose and the object of the project; the development of the project and the development of the project itself. These were the ISO 9001: 2015 standards on quality management, the ISO 21500: 2012 standards on project management guidelines and the ISI 45001: 2018 standards on risk management. They ensure the sustainability of the actions envisaged but also the significant improvement of the results. Following the analysis of the coherence of the SCDOGa/HO1 project, we had to proceed to the clarification of several phases or stages of the project, by establishing the "project document".

2.3 Project clarification: The SCDOGa/HO1 project document

a) Presentation of the project

- ***Description of the project area***

The study was carried out in the city of Franceville, capital of the Haut-Ogooué province, precisely in the Mpassa department (Fig 1). The city is composed of a large mix of ethnic cultures. According to the latest 2015 General Census of Population and Housing (RGPL), the commune of Franceville has approximately 110,568 inhabitants (DGS-RGPL 2013, 2015).

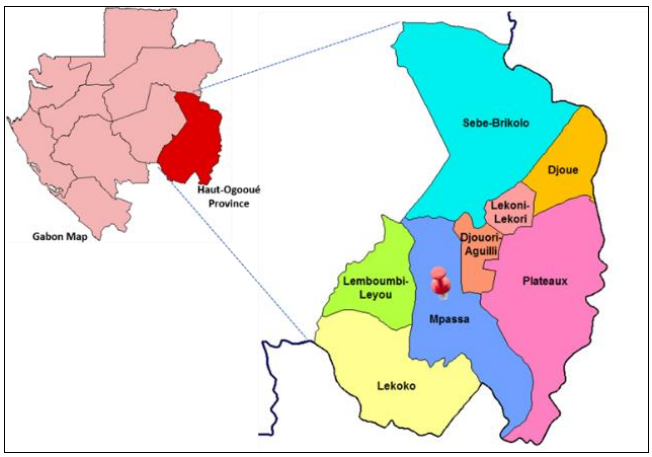

Fig 1: Overview of the province of Haut-Ogooué in a map of Gabon and Administrative division of the province of Haut-Ogooué into departments

- ***Presentation of the project leader or promoter***

In the case of the SCDOGa/HO1 project, the project sponsor is a Gabonese non-governmental organization (NGO). It aims to support education, research, and care for patients with sickle cell disease and other hemoglobinopathies in Gabon.

- ***Background of the project***

The lack of knowledge or ignorance of sickle cell disease by the populations in Africa in general and in Gabon in particular, are reasons that could explain the high rate of the disease in the continent. Sickle cell disease is the most common genetic disease in the world. This mobilization will contribute to the improvement of the follow-up and management of sickle cell disease patients throughout the country.

- ***Identification of the problem***

First of all, we had to clearly define the problem of our study and for this we used the QQOQCP (What, Who, Where, When, How and Why) quality analysis tool with the aim of identifying the problem of our study from all its angles and in a complete manner.

b) Project environment

- ***Analysis of the real expectations of the target population***

Although some patients are still unaware of the dark nature of this disease, because the patient is not cured in Africa, the fact remains that for other patients, the best hope to live longer with this disease would be to benefit from a follow-up and a global medical care. Therefore, obtaining the long-term Disability (LTD) card and its use in medical facilities on the one hand, and the assignment of a referral doctor for follow-up on the other hand, are the real expectations of the target population of the SCDOGa/HO1 project.

After clarifying the expectations of the project's target population, we conducted a SWOT analysis that aimed to highlight the strengths, weaknesses, opportunities, and threats of the SCDOGa/HO1 project's supporting structure.

- ***Technical study of the project***

The technical study of the project consisted in planning, organizing and controlling all the means likely to be mobilized before, during and after the implementation of the project. This included human, financial and material resources, as well as communication means and partnership contracts. This step would allow the effective and efficient implementation of all the activities of the SCDOGa/HO1 project.

- ***Economic model of the SCDOGa/HO1 project***

This is undoubtedly the longest and most complex point of the project document in view of the number of steps it includes. It allowed us to verify that the project leader did not omit any important points when writing the project document. The project's business model includes the following steps: beneficiary segment, value proposition, relationship with beneficiaries, distribution channels, key activities, key resources, key partners and cost structuring.

- ***Planning the monitoring and evaluation process***

This is a major step that allowed us to complete our evaluation process. It consisted of determining the various steps and activities planned during the evaluation process of the SCDOGa/HO1 project. The main lines of the evaluation process of this project are presented in Fig 2.

2.4 Sampling and Data Recording

a) Questionnaire and enrolment

The form was designed for families with at least one member with sickle cell disease. We defined a set of questions to know the family context of the respondent. These questions varied according to the respondent. In fact, since our form is conditional, its size changes according to the answers to certain questions (case of cascade questions). The responses made up the 79 columns of our CSV database file. During the study period, 2778 participants were interviewed and constituted the 2778 rows of our CSV database file. Interviews were conducted in Gabon, in 1

province, 5 departments, 5 communes, 6 arrondissements, and 77 neighbourhoods. one entry had too much missing data and was deleted, a total of 2777 entries will be analysed throughout the study. The teams were deployed in the field in variable groupings according to the number of people present and available. Equipped with white and green, fluorescent T-shirts and vests respectively. The field investigators went to meet the families in the different neighbourhoods, with the form in paper format. These results were then brought back to the NGO's headquarters in Franceville (Gabon), to record the paper results on the digital form (followed by a record in the database). 6 interviewers at headquarters did the same enrolment work at headquarters and recorded other respondents directly via the digital form (followed by recording in the database). Homes with at least one parent or adult child were included in the study. All closed homes and those where the caretakers were not present were not interviewed.

b) Questionnaire and Scanning

Considering the postulate that error is human, we must do our utmost to reduce it because the presence of errors can partially or significantly distort the statistical results. Therefore, the data collection step is often essential and must be done with care.

We have created a digital form, a partial view of which is shown in Fig 3, which is used to record respondents directly at headquarters or off-line via a previously completed paper form. Thanks to the digital form, errors in the format of the dates were limited with the appearance of a virtual calendar during data entry. We have listed most of the provinces, departments, and communes of Gabon. Therefore, selecting a province from a drop-down list will update the available department choices. Selecting a department also updates the related communes. This is an example of the benefits of digitization, knowing that it has also been applied for example to the age of parents, the selection of family type or ethnicity. At the end of entering all the answers of a respondent, we can make a record in the database with one click. In case of multiple errors, it is possible to delete all fields. When several computers are recording independently, all database CSV files can be placed in a common folder and combined in one click to create a single database CSV file.

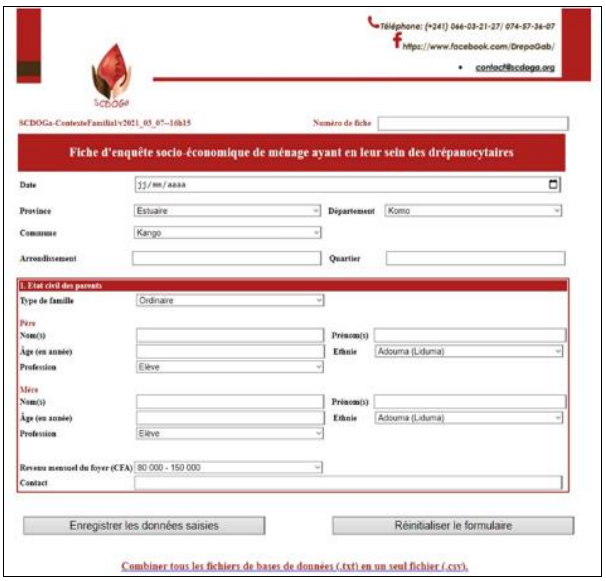

Fig 2: Partial digital form

This form was developed in the format of a website programmed in php. Fig 4 shows the tree structure of the files and folders necessary for the functioning of the website representing the digital questionnaire. Everything was developed in php and the website runs on a fixed computer disconnected from the internet thanks to a local server (WampServer in the case of the NGO SCDOGa). This website was created by the data analysis department of our NGO. It can easily be put on an online server, for a national or international data collection campaign, if the interviewers have a permanent or semi-permanent internet connection on devices used for enrolment (phone, tablet, minicomputer, connected watch). The index.php file and the images folder are the basic elements for the proper use of the web page (home page) of the form in Fig 3. The javascript folder gathers the functions managing the display of the questions in cascade. Let's take the example of the question "Did you know about sickle cell disease?" Answering Yes will be followed by the appearance of the cascading question "Have you ever been screened?". Answering No, will display the other cascading answer "Why?". The functions for saving data, deleting form fields, or combining different database files are contained in the php folder. The bdd_csv folder contains a single database csv file. The bdd_txt folder will contain the txt file representing the global database obtained from all the csv files in the bdd_csv folder. This is useful when the database csv files from all the other computers used for data collection are brought together on the same computer with the aim of combining them into a single file. Finally, the readme file is an explanatory note for all interviewers. Its main purpose is to help the interviewers to quickly get used to the digital form.

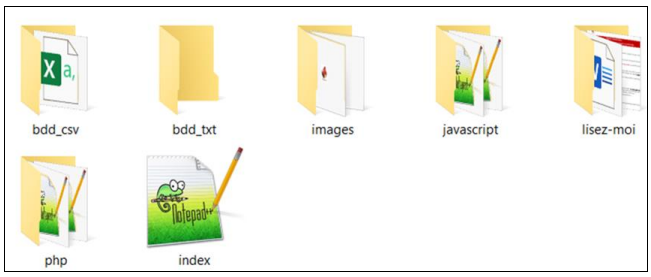

Fig 3: Files and folders necessary for the operation of the digital questionnaire

2.5 Statistical analysis

We analysed the data with R software (version 4.2.1) and its environment RStudio (version 1.1.445). The qualitative data were described in percentages and the quantitative data in means and standard deviation. For the bivariate analysis (two by two) of the variables, the means were compared with the student test; the qualitative variables with the chi-2 and Fisher test. The accepted significance level was $\alpha=0.05$.

3. Results and discussion

3.1 Sample Description

The survey was conducted in the department of Mpassa, precisely in the four districts of the commune of Franceville, over a period of six months from December 8, 2021 to June 8, 2022. The teams were deployed in the field from Monday to Friday during the entire survey period. A total of 2777 households in 55 neighbourhoods responded to our questionnaire. The communes of the 1st, 2nd, 3rd and 4th districts were those concerned by the study. Fig 4 shows the

proportion of the population that participated in the study within each arrondissement of the commune of Franceville. 52,2% of the households surveyed were ordinary households (father, mother and children), 32% were single-parent households, 14% were extended families and 2% were blended families (Fig 5).

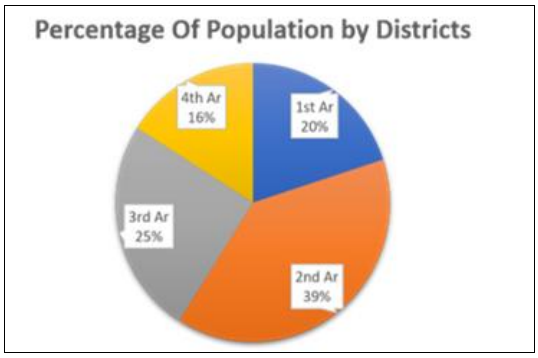

Fig 4: Percentage of population by districts

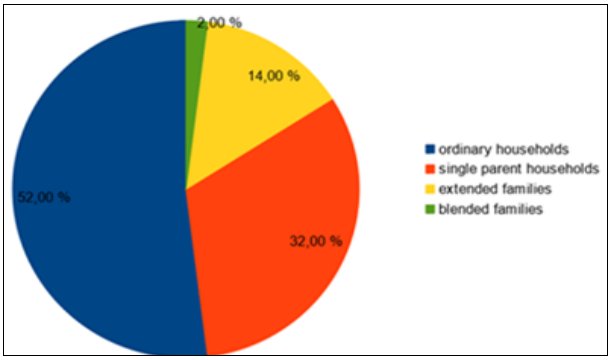

Fig 5: Percentage of distribution by family type population

Almost all ethnic groups were represented; in 61% of the cases both members of the couple were of the same ethnic group and in 39% of the cases the members of the couple were of different ethnic groups.

Regarding the parent's occupation, a majority are in the private and high school sectors. However, in the private sector, there are more men (23.8%) than women (15.7%). Percentages in the high school sector are the same between men and women. Few parents are university students (1.65%) or unemployed (2.95%). Civil servants, on the other hand, were predominantly represented by men (9.5%).

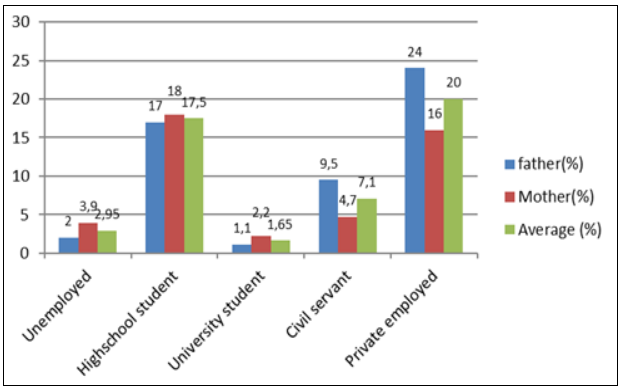

Fig 6: Activities practiced by the father and mother

3.2 Knowledge of sickle cell disease and sickle cell status of parents

The question concerning knowledge of sickle cell disease was asked in the households. We note that Sickle cell disease in the study population was found to be a known disease. However, there is a significant difference between knowledge in mothers and fathers in favour of mothers. 1505 (54%) of the fathers do not know this pathology for 46% who do. In this total of 2777 households only, 1313 (47.28%) fathers answered the question on the effectiveness of their screening (Table 1).

Table 1: Knowledge of fathers vs. knowledge of mothers

		Mother			
		No	Yes	Total	p-value[1]
Father	No	452 (30%)	1 053 (70%)	1 505 (100%)	<0,001
	Yes	473 (37%)	799 (63%)	1 272 (100%)	
	Total	925 (33%)	1 852 (67%)	2 777 (100%)	

[1]chi-square test for independence

Overall, 72% (940) of these fathers had never been screened for sickle cell disease (hemoglobin electrophoresis) compared to 373 (28%) who had been screened.

We note that for those who had performed their screening tests 81% were HbAA for 18% carriers of the sickle cell gene HbAS/HbAC, the rest had forgotten their hemoglobin status (Table 2).

Table 1: Carrying out the screening

	Have You Ever Been Screened? Mother				
	No	Yes	Unknown	Total	p-value[1]
Have You Ever Been Screened? Father					<0,001
No	447 (48%)	99 (11%)	394 (42%)	940 (100%)	
Yes	45 (12%)	246 (66%)	82 (22%)	373 (100%)	
Unknown	685 (47%)	345 (24%)	434 (30%)	1 464 (100%)	
Total	1 177 (42%)	690 (25%)	910 (33%)	2 777 (100%)	

[1] chi-square test for independence

The same analysis was performed in the mothers, 67% of the mothers knew the disease and 37% did not (Table 1). 910 of the 2777 mothers did not wish to answer the question on the effectiveness of their hemoglobin electrophoresis test; among the mothers from the other households, 1177 (63%) had never performed the test, compared to 690 (37%) who had performed their test. It is noted that for those who did perform their screening tests 80% were HbAA for 19% carriers of the sickle cell gene HbAS/HbAC, 1.2% had a hemoglobin status HbSS/HbSC and the rest had forgotten their hemoglobin status (Table 2)

According to Table 1, there is a significant difference p-value<0.001, mothers have a better knowledge of the disease than fathers.

The results presented here show a significant difference in the level of screening for sickle cell disease and that more mothers were screened. The main reasons for not screening fathers and mothers are mainly lack of communication, lack of interest, and lack of education about this disease to varying degrees (Table 3).

Table 2: Main reasons for not screening

Why Father	Why Mother					
	Lack of education	Lack of interest	Lack of communication	Unknown	Total	p-value[1]
Lack of education	41 (17%)	17 (7,1%)	14 (5,9%)	167 (70%)	239 (100%)	<0,001
Lack of interest	27 (4,5%)	74 (12%)	33 (5,4%)	472 (78%)	606 (100%)	
Lack of communication	18 (2,7%)	13 (1,9%)	240 (36%)	402 (60%)	673 (100%)	
Unknown	79 (6,3%)	171 (14%)	221 (18%)	788 (63%)	1 259 (100%)	
Total	165 (5,9%)	275 (9,9%)	508 (18%)	1 829 (66%)	2 777 (100%)	

In order to see the level of genetic mixing of the hemoglobin status of the couples, we made crosses according to the hemoglobin status of the two members of the couple (Table 4).

Table 3: Father's status vs. mother's status

	hemoglobin status of the mother						
	AA	AS/AC	NSP	SS/SC	Unknown	Total	p-value
hemoglobin status of the father							
AA	169 (56%)	27 (8,9%)	0 (0%)	1 (0,3%)	105 (35%)	302 (100%)	
AS/AC	21 (30%)	28 (41%)	0 (0%)	0 (0%)	20 (29%)	69 (100%)	
NSP	0 (0%)	0 (0%)	0 (0%)	0 (0%)	2 (100%)	2 (100%)	
Unknown	359 (15%)	77 (3,2%)	1 (<0,1%)	7 (0,3%)	1 960 (82%)	2 404 (100%)	
Total	549 (20%)	132 (4,8%)	1 (<0,1%)	8 (0,3%)	2 087 (75%)	2 777 (100%)	

Mothers and fathers with sickle cell trait or sickle cell disease enter relationships regardless of their partners' unknown hemoglobin status (3.2% and 0.3%) or knowing their problematic HbAS, HbAC, HbSS or HbSC status (41% and 29%) Table 5.

Table 4: Number of children screened by father's status A and B Number of children screened by mother's status

A	father's hemoglobin status					
	AA	AS/AC	NSP	Unknown	Total	p-value
How many Children Screened						
Some Children	9 (24%)	5 (13%)	0 (0%)	24 (63%)	38 (100%)	
All Children	44 (30%)	17 (11%)	0 (0%)	87 (59%)	148 (100%)	
Unknown	249 (9,6%)	47 (1,8%)	2 (<0,1%)	2 293 (88%)	2 591 (100%)	
Total	302 (11%)	69 (2,5%)	2 (<0,1%)	2 404 (87%)	2 777 (100%)	

B	Mother's hemoglobin status						
	AA	AS/AC	NSP	SS/SC	Unknown	Total	p-value
How many Children Screened							
Some Children	13 (34%)	13 (34%)	0 (0%)	1 (2,6%)	11 (29%)	38 (100%)	
All Children	66 (45%)	33 (22%)	0 (0%)	6 (4,1%)	43 (29%)	148 (100%)	
Unknown	470 (18%)	86 (3,3%)	1 (<0,1%)	1 (<0,1%)	2 033 (78%)	2 591 (100%)	
Total	549 (20%)	132 (4,8%)	1 (<0,1%)	8 (0,3%)	2 087 (75%)	2 777 (100%)	

It is to be regretted that mothers screen more children than fathers and mothers with SS or SC status screen less than other mothers, 2.6% of the latter screen some of their children while 4.1% screen all their children. Mothers with heterozygous status screen the same proportion of their children (34%) as mothers with AA status. In general, this table shows that mothers who do not have an abnormal hemoglobin status screen more of their children (45%).

3.3 Number of children screened by monthly income

a) Screening performed regardless of age

To evaluate the level of income of the households surveyed and their situation in relation to sickle cell status, we asked the respondents about the amount of their monthly income. 2,671 households out of the total sample, or 96%, had a monthly income of between 80,000 and 150,000 FCFA (Fig 7).

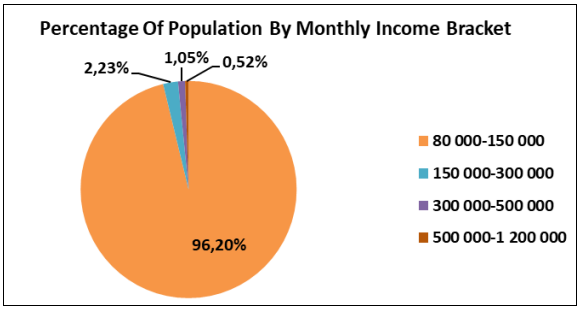

Fig 7: Distribution of The Surveyed Population According to Monthly Income

In order to evaluate their situation in relation to sickle cell disease status with the level of income of the households surveyed, we asked the respondents about the effectiveness of screening for sickle cell disease in their various households at different times in their children's lives (screening at birth or later) see result in Table 6 and 7.

Table 5: Number of children screened by monthly income levels

	Monthly Revenue						
	150 000 - 300 000	300 000 - 500 000	500 000 - 1 200 000	80 000 - 150 000	Unknown	Total	p-value[1]
How many Children Screened							<0,001
Some Children	0 (0%)	0 (0%)	0 (0%)	38 (100%)	0 (0%)	38 (100%)	
All Children	9 (6,1%)	5 (3,4%)	5 (3,4%)	129 (87%)	0 (0%)	148 (100%)	
Unknown	53 (2,0%)	24 (0,9%)	9 (0,3%)	2 504 (97%)	1 (<0,1%)	2 591 (100%)	
Total	62 (2,2%)	29 (1,0%)	14 (0,5%)	2 671 (96%)	1 (<0,1%)	2 777 (100%)	

[1] Fisher's exact test

Households with the lowest incomes (80,000 to 150,000 FCFA) screen more of their children, with 87% (Table 6) screening all their children. If income conditions screening, is there a link between the number of annual seizures and income?

b) Income-based birth screening

A total of 28 of the 160 households that agreed to answer this question had performed the neonatal sickle cell test on their children. The number of children screened at birth appears to be associated with income, but this is not very significant, Table 7.

Table 6: Neonatal screening based on monthly income levels

	Monthly Revenue						
	150 000 - 300 000	300 000 - 500 000	500 000 - 1 200 000	80 000 - 150 000	Unknown	Total	p-value[1]
Child Birth Test							<0,001
No	9 (5,6%)	4 (2,5%)	4 (2,5%)	143 (89%)	0 (0%)	160 (100%)	
Yes	0 (0%)	1 (3,6%)	1 (3,6%)	26 (93%)	0 (0%)	28 (100%)	
Unknown	53 (2,0%)	24 (0,9%)	9 (0,3%)	2 502 (97%)	1 (<0,1%)	2 589 (100%)	
Total	62 (2,2%)	29 (1,0%)	14 (0,5%)	2 671 (96%)	1 (<0,1%)	2 777 (100%)	

[1] Fisher's exact test

3.4 Discussion of Findings

This study involved conducting a socioeconomic survey to identify these parameters in households that may have one or more children with sickle cell disease. Although it is higher than in high-income countries and is affected by non-specific socioeconomic factors, the estimated mortality among children with sickle cell disease.

Sickle cell disease in Franceville remains a partially known disease. The lack of knowledge in this region is justified by a lack of communication. Indeed, as a marginalized disease, it remains a taboo among the Gabonese population. The latter confuse it with a spell and do not dare to talk about it for fear of being excluded because of ignorance and prejudice related to this disease. Similarly, according to Doris Bonnet in 2000 [17], sickle cell disease is thought to be a manifestation of the possession of the sick child and his family by a sorcerer or by an evil spirit [17]. Moreover, the awareness campaigns that are carried out in Gabon about sickle cell disease are not sufficiently relayed by the media, which hampers the expected impact. Women are more knowledgeable than men on this subject. This could be explained by the fact that, on the one hand, screening for the disease is recommended to them during the prenatal check-up and, on the other hand, that they are the most likely partner in case of disease in their child. These results corroborate those found by Tsala in 2009, which show that in Sub-Saharan Africa, the mother is designated as responsible for this transgression [18].

In the total population studied, only 28.77% were in couples with knowledge of sickle cell disease, 16.27% were in couples who were unaware of the existence of the disease, and the rest were in relationships where at least one or both partners were unaware of the disease. This way of choosing a partner regardless of his or her hemoglobin status. When HbS is inherited from only one parent, the heterozygous (HbA/HbS) child is usually an asymptomatic carrier, although some symptoms may be present depending on the expression level of each allele [19]. Indeed, the effect of carrying the sickle cell trait is not written on a person's physique, so it is very difficult in the context of choosing a partner to make an informed choice without a screening test.

Screening for sickle cell disease is done by an examination called hemoglobin electrophoresis test. 38.28% (373 men and 690 women) of the total population studied have a known status. This could be justified on the one hand, by the fact that the latter only see this screening as an additional expense not covered by insurance and which they could well do without, especially since they do not present any symptoms. On the other hand, by the fact that some people, although having knowledge of the disease, do not submit to the examination for fear of the consequences of its existence. The same observation was made by Doris Bonnet in Côte d'Ivoire in 2001 [20]. Screening for sickle cell disease is prescribed by doctors to pregnant women, but the latter very rarely undergo it. As a result, carriers are often not informed of their condition until after the birth of a sickle cell child. As screening at birth is not mandatory in Gabon, it is only after a series of infections or several anemia attacks that the status of the sick child is discovered. From a jurisdictional point of view, this examination is recommended for couples wishing to marry in the list of premarital examinations, but there is no law prohibiting the marriage of two individuals with the condition heterozygous hemoglobin status.

Individuals with a known status are predominantly normal homozygous HbAA from a clinical point of view. However, many households are made up of two heterozygotes (HbAS). Knowing one's electrophoretic status is not an obstacle in the formulation of couples in this region. As part of its neotality policy, Gabon promotes births by setting up actions in favor of new mothers. In our malaria context, where children with sickle cell disease die before the age of 5 years due to lack of adequate care [21], it is risky to adopt this kind of behavior. Indeed, in the cultural context, the death of a child is never natural and therefore almost never linked to this kind of pathology which is often not detected or known before the death of the child [22].

The results obtained show a predominantly poor population (96%) with a monthly income below 150,000 XFA. The category with this type of income is the one with the most children screened. However, in this context, screening is not directly related to social category because this class alone constitutes almost the entire study population. In addition, those with a monthly income of more than 150,000 XFA screen all their children and do not have more than one sickle cell child. This category of households not only has the most children but also the most children screened at birth. One might think that this attitude of the parents would be due to knowledge, but in fact, it is the warning signs of recurrent in the event of illness and hospitalizations in 56.1% of cases, anemia and repeated pain observed in the children concerned. In cases where one parent knows his or her status, the couple will tend to do a neonatal screening for sickle cell disease, especially when it is the mother's status that is known. Indeed, the lack of communication about the disease, the lack of interest and lack of education about the disease do not contribute to a systematic and voluntary screening on the part of the couple concerned. For 3.7% of the population, the death of a close relative during a sickle cell crisis was the point at which they became aware of the seriousness of this pathology and therefore wanted to perform hemoglobin electrophoresis on all or some of the children in the household.

The individuals who are aware of it are represented by the female population to the detriment of the male population. The rate of screening carried out in the said population is significantly lower (26%) than the absence of screening (74%). As a result, most of the households encountered had unknown status. Those who knew their status were mainly AA homozygous. The population of Franceville was found to be poor overall, with a monthly income of less than 150,000. However, there was no correlation between monthly income and sickle cell disease screening and between monthly income and having a sickle cell child. 2389 (89%) adults had children in their homes but only 187 (7.8%) had them screened. Screening occurred at the time of symptom onset. Only 28 adults screened their children at birth. 44 children with sickle cell disease were identified during the study. Indeed, 36 (81%) children, ranging from kindergarten to university, are enrolled in school. However, it is not a subject that is easily discussed because the academic environment of these children is not necessarily informed of their health status. In this panel of children with sickle cell disease, 35 (79%) of them are taken care of by through the conventional CNAMGS system. Of course, this care reduces the costs of medication and hospitalization, but it is not enough to alleviate the suffering inflicted by this disease. Indeed, the medical care of a sickle cell child can go up to 1.000.000 XFA or 1.500.000 XFA per week on

average. How can these families live, send their children to school, and take care of their sickle cell children's health, even if only one child among the siblings suffers from this pathology? Support in medical care is often the main request of parents. There is a special system of the CNAMGS aiming at supporting the medical care of people living with a long-term disease but these people have no knowledge on this special advantage set up by the Gabonese state. It is therefore important, in view of the above, that information operations on this pathology and on the means of care be put in place to avoid cases of sickle cell disease in our territory.

4. Conclusion

Finally, sickle cell disease is not very well known in the commune of Franceville. The lack of communication about this disease and the confusion between sickle cell status and blood type could be the reason for this. Screening for sickle cell disease is not included in the habits of the population, only 26% of the study population has already been screened only 7.8% of households screened their children. The population of Franceville was found to be poor overall, with a monthly income of less than 150,000. However, there was no correlation between monthly income and sickle cell disease screening and between monthly income and having a sickle cell child. Low income could negatively influence the medical management of a person living with sickle cell disease.

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