Acta Scientific MEDICAL SCIENCES (ISSN: 2582-0931)

Volume 7 Issue 11 November 2023

Research Article

Prevalence of Sickle Cell Disease Among Undergraduate Students of the University of Bamenda, North West Region of Cameroon

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DOI: 10.31080/ASMS.2023.07.1701

Received: September 13, 2023

Published: October 20, 2023

Abstract

Background/Aims: The term sickle cell disease (SCD) refers to a collection of hemoglobinopathies (inherited blood disorders) characterized by abnormal hemoglobin and produced through the homozygous inheritance of a sickle cell allele. Heterozygous inheritance results in a condition known as sickle cell trait (SCT). Individuals with sickle cell trait have a 50% chance of passing the gene to future offspring. The sickle cell trait is also known to be associated with several rare but serious complications including renal complications, renal cancer, spleen damage, and exercise-related sudden death. We therefore aimed to determine the prevalence of sickle cell trait and sickle cell disease amongst students attending the University of Bamenda and to assess their knowledge, attitudes and practices towards the disease.

Methods: This was a cross-sectional study involving students of the University of Bamenda, with blood samples collected and analyzed (Hb Electrophoresis) at the Bamenda Regional Hospital between April and June, 2020. Questionnaires were used to obtain socio-demographic data.

Results: There were 250 participants. Forty (16%) participants had the sickle cell trait and no participant (0%) was positive for sickle cell disease. Of the 40 participants with SCT, 14 (5.6%) were males and 26 (10.4%) females. Thus, majority (84%) of the participants carried the normal gene (AA). Most students (> 90%) had knowledge of sickle cell trait and disease and of the opinion to consider genetic counseling and testing before marriage as the appropriate preventive measure to control the disease in the society.

Conclusion: Sickle cell disease amongst students of University of Bamenda is rare. However, more than 10% of the students had the sickle cell trait, underscoring the importance of disease sensitization and genetic screening to curb the burden of SCD in this region of Cameroon.

Keywords: Genetic Counselling; Newborn Screening; Sickle Cell Disease; Sickle Cell Trait; Haemoglobin Electrophoresis

Abbreviations

SCT: Sickle Cell Trait; SCD: Sickle Cell Disease

Introduction

The term sickle cell disease (SCD) refers to a collection of haemoglobinopathies (inherited blood disorders) characterized by abnormal hemoglobin and produced through the homozygous inheritance of a sickle cell allele. Heterozygous inheritance results in a condition known as sickle cell trait (SCT). Individuals with SCT have a 50% chance of passing the gene to future offspring. In recent years mounting evidence has confirmed that SCT is associated with several rare but serious complications including

renal complications, renal cancer, spleen damage, and exercise-related sudden death [1]. Sickle cell disease is an autosomal recessive genetic disorder; thus, an individual must inherit two recessive genes (i.e., one from each parent) which each code for abnormal beta-globin (HbS) in order for the disease state to occur. If a person receives a normal beta-globin gene (HbA) from one parent and a sickle beta-globin gene (HbS) from the other, that person is said to have sickle cell trait (SCT) or to be a "carrier" of the disease. Individuals with SCT are generally healthy, but have a 50% chance of passing the trait on to their children. As with all autosomal recessive genetic disorders, if each parent has one sickle beta-globin gene and one normal beta-globin gene (HbAS + HbAS), the couple would then have a 25% chance of producing a child with SCD (Hb SS), a 25% chance of producing a child who does not have the disease or trait (HbAA), and a 50% chance of producing a child with SCT (HbAS). If one parent has SCD (HbSS) and the other parent has normal beta-globin (HbAA), then all offspring would have SCT (HbAS). Finally, if a parent has SCD (HbSS) and the other has trait (HbAS), each child would have a 50% chance of having SCT and a 50% chance of having SCD. Individuals with SCT (one normal beta-globin gene and one abnormal sickle gene) generally experience few complications. However, they may pass the sickle cell gene to their own children; thus, knowledge of trait status and education regarding transmission of the gene is critical for individuals with SCT who wish to engage in reproductive planning. Sickle cell anemia has a high prevalence throughout equatorial Africa; additionally, the genetic defect is now known to be widespread in parts of Sicily and southern Italy, northern Greece, southern Turkey, the Middle East, Saudi Arabia, much of central India, and the Americas [2]. The prevalence of sickle cell trait in western, central and eastern Africa varies from 5 to 40% but it is less common in northern and southern Africa [3]. Three quarters of all sickle cell cases occur in Africa and the total number of carriers in the world is estimated to be about 120 million. In Cameroon, the prevalence of the sickle cell trait is estimated to be 18.2% for the heterozygous form and 2–3% for the homozygous SS forms [4]. The prevalence of SCD is increasing and becoming very common in Cameroon. Sickle cell trait is generally a benign medical condition, and most individuals with SCT are healthy and have a typical lifespan. As a result, a person with SCT may be unaware of their positive trait status [5,6]. However, an individual with SCT has a 50% chance of passing on the abnormal sickle cell gene to any future offspring, making knowledge of trait status critical if the individual desires to engage in informed reproductive decision making [5-10]. Early knowledge of sickle cell status will help students of the University of Bamenda to prevent this disease, since most of them are of the premarital age, some engaged and some in long-term relationships. We therefore aimed to determine the prevalence of SCD and SCT amongst students attending the University of Bamenda, and to assess their knowledge, attitude and practices regarding this disease.

Materials and Methods

Type and duration of the study

This was a cross-sectional study that was carried out between April and June, 2020.

Study site and participants

Study participants were students attending the University of Bamenda**.** Blood samples were collected from students on campus, temporally stored in a cold-chain and transported same day to the Haematology Unit of the Bamenda Regional Hospital for Hb Electrophoresis analysis. The University of Bamenda is located in Bambili, Tubah Subdivision, North West Region of Cameroon.

Ethical consideration

Ethical clearance according to the Declaration of Helsinki was sought from the Institutional Review Board of the Faculty of Health Sciences, University of Bamenda. Free and informed consent was obtained from each participant before inclusion in the study. The right to refuse or withdraw from the study was fully maintained. An authorization was issued by the Director of Regional Hospital Bamenda to analyse samples in their Laboratory.

Data collection

A structured questionnaire was used to collect sociodemographic data, family history of SCD of the participants, and their knowledge, attitude and practices towards the sickle cell disease. Questionnaires were administered in English. Biological data regarding the prevalence of SCT and SCD amongst the students was obtained by laboratory analysis of the blood samples.

Blood collection and biological analysis

A sample of approximately 3 mL of blood was aseptically collected in a tube containing ethylene diamine tetra acetic acid tri potassium (EDTAK3) from students of the University of Bamenda,

and transported at 2 - 8°C to the Bamenda Regional Hospital Laboratory for Hemoglobin Electrophoresis assay.

Tris EDTA Borate (TEB) buffer was used for the qualitative determination of haemoglobin fragments. After centrifugation of the blood tubes, plasma was extracted from the cells. 20µl of the packed red cells were mixed with 380µl of distilled water (1: 20 dilution) and allowed for 20 minutes (lysing). With the power supply disconnected, the electrophoresis tank was prepared by placing equal amounts of Tris EDTA Borate (TEB) buffer (pH 8.6) in each of the outer buffer compartments to a depth of about 2.5cm. Two wet chamber wicks were then placed one along each divider/bridge support ensuring that they made good contact with the buffer. The cellulose acetate paper was soaked by lowering it slowly into a reservoir of buffer and left for about 30 minutes prior to use. A drop each of the control and test haemolysates was placed accordingly on the well-plate. The cellulose acetate strip was removed from the buffer and blotted twice between two layers of clean blotting paper to remove excess buffer but not allowed to dry. By means of an applicator, the control and test haemolysates were applied on the cellulose acetate membrane and carefully introduced onto the frame of the electrophoretic tank, with both ends in contact with the buffer. The lid of the tank was replaced and the tank connected to a power supply of 250 volts and current 50mA and allowed to run for 20 minutes. The power supply was then disconnected, the membrane removed and results obtained by comparing test samples with those of controls (haemolysates from a known sickle cell trait sample). Results were later distributed to students and genetic counseling was done.

Data management and analysis

Results were recorded into a log book. Completed questionnaires were coded and data keyed into Microsoft Office Excel and stored in the laptop and flash drives which minimized the chances of losing data. Prevalence of SCT and SCD were calculated from the proportion of positive samples. We analyzed data using the Statistical Package for Social Sciences (SPSS).

Results and Discussion

Sociodemographic characteristics of study participants

A total of 250 students participated in the study; 104 (41.6%) males and 146 (58.4%) females. The preponderance of females in this study is similar to that obtained by Ngwengi., *et al.* [13]. Majority (50.8%) of the students were aged 16-20 years, while those within the age groups of 21-25 years, 26-30 years, and >30 years constituted 38.8%, 8%, and 2.4% respectively. Most (94.4%) of the participants were single, while 5.6% were married (Table 1). A similar study involving undergraduate students of the University of Calabar by Valerie., *et al.* [14] had socio-demographic characteristics similar to those obtained in this study.

Variable	Category	Frequency	Percent (%)
Gender	Female	146	58.4
	Male	104	41.6
Age range	16-20 Years	127	50.8
	21-25 Years	97	38.8
	26-30 Years	20	8
	>30 Years	6	2.4
Marital Status	Single	236	94.4
	Married	14	5.6

Table 1: Socio-demographic Characteristics of Students attending the University of Bamenda.

Prevalence of SCT and SCD

The prevalence of SCT and SCD amongst students attending the University of Bamenda was 16% and 0% respectively. Thus, 84% of the population had the normal gene AA (Figure 1). The prevalence recorded in our study was less than those reported in a similar study conducted in Cameroon of 18.2% and 2-3% for SCT and SCD respectively [4]. The 0% prevalence of SCD amongst students in the University of Bamenda could be due to the fact that most persons with SCD have a shortened life span. They usually suffer from sickle cell disease crisis and hospitalized [2,5], and this may affect their studying to higher levels.

Prevalence of SCT and SCD according to gender

There were more females than males with SCT but this difference was not statistically significant ($p > 0.05$) (Table 2). This is in agreement with the findings of Ngwengi., *et al.* [13] who recorded an insignificant higher prevalence of sickle cell trait among females compared to males. Given the female preponderance of our study cohort, we may suggest from our findings that the Hb status of an individual is not influenced by gender.

	Prevalence of Sickle cell disease and sickle cell trait		Total	P. value	X^2
	AS	AA			
Gender					
Female	26	120	146	0.36	0.854
Male	14	90	104		

Table 2: Prevalence of SCD and SCT according to Gender.

Prevalence of SCD/SCT according to family history of SCD

We recorded an association between family history and the prevalence of sickle cell disease/trait (p-value < 0.01, at Confidence interval 95%); with the trait being more prevalent (38.3% versus 10.8%) in those who reported having a family history of the disease (Table 3). This confirms the fact that SCD is an inherited blood disorder.

	Category	Family History of sickle cell disease		Total	P-value (CI = 95%)
		No	Yes		
Prevalence of Sickle cell disease and sickle cell trait	AS	22	18	40	<0.01
	AA	181	29	210	
Total		203	47	250	

Table 3: Prevalence of sickle cell disease or sickle cell trait with respect to family history of the disease.

Distribution of Respondents with respect to knowledge regarding SCD

Awareness of the disease

Majority (92%) of the respondents were aware of the sickle cell disease and traits. Majority (95.2%) were also aware it is an inherited blood disorder, while 4.4% were of the opinion that SCD is an infectious disease and 0.4% of respondents considered SCD to be a fatal illness (Table 4). Most of the participants were aware of the genetic transmission of sickle cell genes which was higher as compared to the study by Boadu., *et al.* in Nigeria where only 48.3% of subjects were aware of the genetic transmission [12]. This could be because our study involved participants of a higher institution of learning.

Do You know about sickle cell disease?	Frequency	Percentage
No	20	8
Yes	230	92
Total	250	100
What is sickle cell disease (SCD)?		
Inherited blood disorder	238	95.2
Infectious disease	11	4.4
Fatal illness	1	0.4
Total	250	100

Table 4: Distribution of respondents based on their knowledge about sickle cell disease.

Knowledge on cure and management of SCD

Concerning knowledge regarding the management or cure of the disease, majority (62.8%) of the respondents were of the opinion that sickle cell disease cannot be cured; 22.8% did not know whether it can be cured or not, and 14.4% of respondents believed that SCD can be cured. These records are slightly lower when compared to those of the study by GN Bazuaye., *et al.* where 15.1% of subjects believed it can be cured, 24% had no idea; while 60.9% (slightly lower than our data) believed it is incurable [11]. Majority (44%) of the respondents said SCD can be managed via genetic counseling; 40.4% were of the opinion that SCD can be managed by use of preventive medicines, while others proposed the use of Vitamins (10%) and Exercise (1.6%) (Table 5).

Do you think there is a cure for sickle cell disease	Frequency	Percentage (%)
No	157	62.8
Yes	36	14.4
I don't know	57	22.8
Total	250	100
How to manage Sickle cell disease		
Preventive Medicine	104	40.4
Vitamins	29	10
Genetic counseling	114	44
Exercise	2	1.6

Table 5: Distribution of respondents based on their knowledge regarding the management and cure of sickle cell disease.

Knowledge regarding estimated life expectancy for SCD patients

Majority (64.4%) of study participants said the estimated life expectancy was 20-30 years followed by 22.4% who said it's between 31-40. Only 4.4% mentioned the estimated life expectancy could be 51 years and above (Table 6). It is common believe that individuals with SCD have a shorter lifespan; thus, most respondents were prone to choosing the lowest age range that was provided in the study.

Estimated life expectancy (Years)	Frequency	Percentage (%)
20-30	161	64.4
31-40	56	22.4
41-50	22	8.8
51 and above	11	4.4
Total	250	100

Table 6: Distribution of respondents based on their knowledge concerning estimated life expectancy of someone with sickle cell disease.

Knowledge regarding disease prevention

Majority (79.2%) of our study participants indicated having knowledge regarding the prevention of SCD in the society, and most (70%) suggested genetic screening before marriage as the appropriate measure for disease prevention (Table 7). This awareness may be the achievement of sensitization programs in the country on SCD/SCT and the fact that respondents were from a higher learning institution.

Knowledge of preventive measures of sickle cell disease in the society	Frequency	Percentage (%)
No	52	20.8
Yes	198	79.2
If Yes, specify		
Genetic counseling	75	30.0
Genetic screening before marriage	195	70.0
Total	250	100

Table 7: Distribution of respondents based on knowledge of preventing sickle cell disease in the society.

Distribution of Respondents according to Attitudes and Practices towards SCD

Stigmatization of SCD patients

As concerned stigmatization of persons with SCD, 99.6% of the respondents considered sickle cell disease to be a disease like any other, while 0.4% of the respondents could stigmatize persons with sickle cell disease through name-calling and regarding the disease as a possible punishment from God (Table 8). This percentage (0.4%) was lower compared to records from previous study by GN Bazuaye., *et al.* in Nigeria, where 18% of the secondary level students will stigmatize persons with SCD [11]. This could be because our study involved university students who are more enlightened and mature.

Stigmatization of patients with sickle cell disease	Frequency	Percentage (%)
No	249	99.6
Yes	1	0.4
Total	250	100

Table 8: Distribution of respondents based on whether or not they used to stigmatize persons with sickle cell disease.

Attitudes regarding the management of sickle cell disease crisis

Almost all the respondents (98.8%) would prefer consulting a physician in case of any crisis related to SCD, while 0.8% and 0.4% of the respondents would prefer to visit a chemist or a traditional healer respectively (Table 9). Our findings suggest that at university level, students are well educated and sensitized regarding the management of this disease.

Whom to visit in case of SCD crisis	Frequency	Percentage
Traditional healer	1	0.4
Chemist	2	0.8
Physician at the Hospital	247	98.8
Total	250	100

Table 9: Distribution of respondents based on their attitudes regarding the management of sickle cell crisis.

Regarding the appropriate timing for an SCD test

Most (72.4%) of the participants were of the opinion that the test for SCD should be performed as soon as a child is born; while others (23.2%) thought this could be done prior to marriage, and very few respondents (4.4%) suggested that the screening for SCD could be added when screening for sexually transmissible diseases (STDs) (Table 10). We suggest based on our findings that newborns be screened for SCD to permit early diagnosis and care, and for parents to participate in sensitizing their children as appropriate to prevent the disease.

Appropriate time for SCD and trait test	Frequency	Percentage (%)
New Born Screening	181	72.4
Health Investigation before Marriage	58	23.2
STD Testing	11	4.4
Total	250	100

Table 10: Distribution of respondents based on their views regarding the appropriate timing for a sickle cell disease test.

Conclusion

In this study, the prevalence of sickle cell trait was 16% while no participant was found to have the sickle cell disease. Majority of the students had knowledge on what sickle cell disease is all about and would consider genetic counselling and testing before marriage as a preventive measure to reduce the burden of sickle cell disease in the society.

Acknowledgements

We thank all participants and the laboratory staff of the Regional Hospital Bamenda for their technical assistance.

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