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"KNOWLEDGE, ATTITUDE AND PRACTICES OF PREMARITAL COUNSELING FOR SICKLE CELL DISEASE (SCD) AMONGST COUNCIL WORKERS IN THE TUBAH, BAMENDA I, II, III AND CITY COUNCIL OF NORTH WEST CAMEROON"

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Abstract: -

Background: Cameroon has high rates of sickle cell disease (SCD) attributed to lack of awareness among the reproductive age in the country. In Cameroon, the prevalence of SCD is 18.2% for the heterozygous form and 2.3% for the homozygous (HbSS) forms.

Objective: This study determined the knowledge, attitude and practices of premarital counseling for sickle cell disease (PMCSCD) amongst Council workers of the Tubah, Bamenda I, II, III and City Council of North West Region of Cameroon, to see how this knowledge can be translated to intending couples to improve marital outcomes.

Materials and method: A community based cross sectional study design with purposive random sampling was used to select Council workers who participated in the study. Data was generated by use of semi-structured questionnaires and presented using frequency, percentages and charts. Descriptive analysis was done using statistical package for social sciences (SPSS) software version 21. Relationship between variables was assessed using Chi-Square Test. We used a *P*-value of <0.05 as a cut off point for statistical significance.

Results: A total of 59 respondents filled the survey. The result shows that knowledge of PMCSCD varied from one aspect to another, with an overall poor level. Attitude of respondents towards PMCSCD remained good (54.2%). Meanwhile implementation of PMCSCD in participants life was grossly inadequate (96.6%), with apparently no premarital counselor for SCD in any of the councils as reported by 83.1% of study participants. Out of the sociodemographic characteristics of interest, level of education was found to be significantly associated with knowledge (X^2*=17.5; P*= 0.04). The survey also found a significant association between participants attitude and sex, age group and marital status. No sociodemographic factor was found to be associated with practice.

Conclusion: Overall, council workers had poor knowledge of PMCSCD. Attitude of workers towards PMCSCD was good. Practice of PMCSCD among study participants remained poor. This suggests that council authorities should take advantage of this information and pull attention to potential areas for intervention and improvements in knowledge of man power resources especially true in relation to sickle cell disease counselor so that the knowledge can be translated to intending couples to foster better outcomes in sickle cell prevention, awareness and management.

Keywords: Sickle cell disease; premarital counseling; knowledge; attitude; practice; awareness.

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INTRODUCTION

1.1. BACKGROUND OF THE STUDY

Sickle cell disease (SCD) is a hematological illness resulting from the inheritance of an anomalous form of hemoglobin [1]. Haemoglobin, a protein found in Red Blood Cells, transports oxygen from the lungs and distributes it throughout the body. Sickle cell disease is caused by a specific mutation where the amino acid Glutamic acid (Glu) at position 6 of the beta chain of hemoglobin is replaced by valine (val) [1-3]. The condition follows an autosomal recessive inheritance pattern and is distinguished by the presence of sickle haemoglobin HbS. The sickle haemoglobin is a genetic mutation of the normal adult hemoglobin HbA, known as either 'SCD SS' or 'sickle cell anaemia' SCA. This mutation is a structural variation induced by a mutation in the haemoglobin gene and is considered the most severe form of the illness [2, 4]. There are more than 700 different types of haemoglobin variations, but the two most significant kinds of haemoglobinopathies that are the most frequent hereditary blood diseases in Africa and worldwide are sickle cell disease and beta thalassemia (expressed as HbS and HbC respectively) [5, 3]. The Sickle Cell Trait, also known as the carrier state, is a condition where an individual has a heterozygous type of the disease. This means that they have up to 40% of the abnormal hemoglobin S (HbS) and do not have anemia [6].

In individuals with sickle cell disease (SCD), the presence of deoxygenated sickled hemoglobin causes the membrane of red blood cells (RBCs) to become deformed, leading to the formation of rigid rods composed of aberrant hemoglobin inside the RBCs. The primary pathophysiological mechanism of sickle cell disease (SCD) is the polymerization of hemoglobin due to an inherited genetic defect. This results in significant alterations in red blood cells. The flexible, spherical biconcave disc-shaped cells of the blood change into hard, sickled – Crescent shaped cells, which further restricts the ability of the haemoglobin molecule to carry oxygen. The atypical red blood cells, which have lost their flexibility and become adhesive and rigid rods, have a tendency to obstruct small blood arteries (such as capillaries and veins). This obstruction leads to a painful episode known as a vasoocclusive crisis, which is a characteristic feature of sickle cell disease. In addition, Sickled Red Blood Cells have a shortened lifetime of 10-20 days, as opposed to the typical lifespan of 90-120 days. The clinical manifestation of sickle cell disease (SCD) is caused by repeated blockage of blood vessels and ongoing destruction of red blood cells. Repetitive blockage of blood vessels causes sudden and long-lasting episodes of discomfort and malfunction in several organs, such as bones, spleen, liver, brain, lungs, kidneys, and joints (resulting in damage to the affected organs). Chronic haemolysis may result in several consequences such as anemia (SCA), gallstones, and jaundice [5, 7]. Sickle Cell Anaemia is a serious outcome of this disorder, but individuals with the sickle cell trait living in areas with high malaria prevalence may have a selective advantage. This implies an evolutionary connection and proposes that genes associated with sickle cell disorder may provide a solution to a cultural problem.

However, those who have a homozygous gene for sickle cell disease HbSS do not get any advantages from this fortification due to their active A-splenia [8].

Chakravorty [4] states that sickle cell anemia is an abandoned chronic illness of increasing global health importance. Patients often experience recurrent illness and hospitalization, resulting in a reduced quality of life. In underdeveloped countries, where healthcare facilities are severely lacking and appropriate treatments are rare, afflicted children seldom live to reach their 5th birthday [5, 9-12].

Ngwengi [5] said that the World Health Organisation (WHO) designated Sickle Cell Disease (SCD) as a Public Health priority because of its significant prevalence.

Approximately 332,000 infants are born globally each year with a significant form of haemoglobinopathy, of which 275,000 are diagnosed with sickle cell disease. Global sickle cell disease (SCD) poses a substantial health burden that is not sufficiently addressed. Approximately 312,000 infants are projected to be born worldwide with sickle cell disease (SCD) each year [13]. The prevalence of this illness is most pronounced in the Sub-Saharan area of Africa (SSA)[13,5].

The sickle cell syndrome was a widely recognized disease in West Africa, known by various local names before its discovery in America in 1910 by Dr. James Herrick, a physician in Chicago. Dr. Herrick first identified elongated shaped hemoglobin in the blood smear of an anemic student from Grenada and used the term "sickle" to describe their crescent shape [14]. In Africa, the death rate among patients aged 5 years and older was between 50% - 90% [15]. In Nigeria, the death rate for children under the age of 5 was over 90% as a result of insufficient and untimely intervention [12].

Globally, SCD is a significant health burden that is inadequately addressed [13].

Individuals living with sickle cell disease face more than only the challenges of managing the severe health consequences of the illness. Sickle cell has caused doubt and discord among families. Due to the unfortunate deaths of many children in the village, families have been compelled to move to designated "safe areas" in order to avoid further harm. These regions are supposed to be free from the perceived curse that afflicted the previous location. This is also the rationale for some names being seen as "bad luck" when translated literally from the vernacular in several African civilizations. Individuals with sickle cell illness often experience stigmatization and discrimination, which may lead families to conceal their affected members [16]. Research has shown a clear connection between having children with this blood illness and a worse quality of life for family members. This is due to the burdens of financial, emotional, and time-related responsibilities [17].

The World Health Organization (WHO) issued a research stating that the carrier frequency of sickle cell disease (SCD) in Cameroon was around 20-30% in the year 2016.Cameroon has the highest annual number of babies with sickle cell disease among Sub-Saharan African nations, with 5 to 18 cases reported.

In Cameroon, as in many other poor countries, there is no comprehensive universal medical health insurance coverage. Cameroon is now undergoing an epidemiologic transformation, characterized by a growing and acknowledged burden of chronic non-communicable illnesses of genetic origin, such as sickle cell anemia (SCA) [19]. The estimated prevalence of Sickle Cell Disease (SCD) in Cameroon is 18.2% for the heterozygous form and 2-3% for the homozygous (HbSS) types [5]. In developing countries, such as those in the third world, the prevalence of sickle cell disease is rising and becoming a significant health concern. In fact, it is estimated that haemoglobinopathies alone in these countries provide a health burden that is similar to the burden posed by other illnesses combined. This is particularly worrying in Cameroon. Providing prenuptial (premarital) counseling services for sickle cell disease (SCD) to screen couples before marriage should be more effective. However, only a small number of couples come for testing since there is little knowledge of SCD, especially among those of reproductive age [21,5].

1.2. PROBLEM STATEMENT

People have ignored premarital counselling and testing for sickle cell disease before marriage, and have based management on palliative care to the sick and the consequences are far too reaching on our communities as evidenced by the ever increasing SCD prevalence in Cameroon but is preventable. Following Law Nº2019/024 of 24 December 2019 instituting the general code of Regional and Local Authorities in Cameroon, local government councils have been charged with responsibilities to include licensing of marriages, births, and deaths, and community health assistance amongst others. Local government councils have so far supported community health efforts only through the provision of equipment and human resources (HCW) to health establishments in their respective areas of command with no intervention strategies to prevent the occurrence of disease (SCD) but could do better. This research was carried out to assess the knowledge, attitudes and practices of council workers towards sickle cell disease, so that we could be able to see how far we could go to sensitize them for premarital counselling and testing for the disease, in order that they could in turn decode this information to develop matrimonial counselling to couple and avoid the consequences of the disease upon the family, society and the nation. As an inherited disease (SCD), the first step towards checking its menace is to prevent its occurrence.

1.3. RESEARCH QUESTIONS

- What is the level of knowledge on premarital counselling and testing for SCD amongst council workers in the Tubah, Bamenda I, II, III and City council of North West Cameroon?
- What are the attitudes of council workers towards premarital counselling for Sickle Cell Disease?
- What practices are involved in premarital counselling for SCD by council workers of the Tubah, Bamenda I, II, III and City Council?

1.4. RESEARCH HYPOTHESIS

HO: Council workers have poor knowledge, attitude and practices towardssickle cell disease premarital counselling.

HA: Council workers have excellent knowledge, attitude and practices towards sickle cell disease premarital counselling.

1.5. RESEARCH OBJECTIVES

1.5.1 General objective

To determine the level of knowledge, attitude and practices of premarital counselling for SCD amongst council workers in the Tubah, Bamenda I, II, III and City councils of North West Region of Cameroon.

1.5.2 SPECIFIC OBJECTIVES

- To assess the knowledge of Council workers on premarital counselling and testing for SCD.
- To identify the attitudes of council workers towards premarital counselling for Sickle Cell Disease.
- To find out the practices involved in premarital counselling for SCD amongst council workers.

1.7. SIGNIFICANCE OF STUDY

This study aimed at establishing the level of information, stance and practice of council workers towards pre-matrimonial counselling and testing for sickle-cell and to see how such knowledge could be translated to couples to improve marital counselling outcomes. The findings from this study shall contribute towards understanding the current SCD-related gaps in knowledge and attitude amongst council workers and will generate Data that will empower policy makers (Mayors) to have some training on prenuptial counselling and testing for SCD. The result of this research will be used to do advocacy to formulate policy through the Assembly to ensure that no Mayor celebrates marriage between any couples whose test results show they are most likely to give birth to sickle celled anaemic children, talk less of carriers. Finally, the result will add to existing literature on the topic for proper management and prevention of SCA and reduction of the cycle of pain, suffering, frustration, suspicion and division that characterises families of people with children suffering from SCA.

1.8. SCOPE AND DELIMITATION OF STUDY

This study is not limited to all council workers who may be involved in registration and or legislation of marriages. The study is limited to all Support Staff of the council.

1.9. DEFINITION OF KEY TERMS

The terms which were used for such study are given below and were defined by [54,86].

- **Genetic disorder:** This is a disorder that is due to any malfunctioning in the DNA sequence of an individual.
- **Haemoglobin:** The carrier protein containing Fe at the centre of it and is the major part of RBCs and allows them to carry O_2 to all parts of the body.
- **Haemoglobinopathy:** A cluster of disorder which causes the synthesis of abnormal molecule of haemoglobin.
- **Sickle cell disease (SCD):** An inherited disorder of the red blood cells in which one gene is for sickle haemoglobin (S) and the other gene is for unusual haemoglobin SC and Thalassemia.
- **Sickle Cell traits:** The inheritance of normal as well as sickle-cell gene by an organism. Children acquire sickle cell from their parents while symptoms don't appear into the parents.
- **Prenuptial counselling for SCD:** Pre- matrimony counselling can be used as an awareness tool for sickle-cell anaemia disease.

- **Haemoglobin A (HbA):** in the composition of Haemoglobin, two alpha & two beta chains are involved, usually synthesized by kids and adults.
- **Sickle cell Anaemia (SCA) SS:** An inherited disorder of the red blood cells in which the haemoglobin is different from the normal haemoglobin. These unusual formed cells are referred as HbSS. These are most severe and common forms of variations of sickle cell disorder.
- **Genetic counselling:** Communication process between health care provider and client that emphasises and provides accurate and up-to-date information about a genetic disorder in a sensitive and supportive and non-directive manner.

LITERATURE REVIEW

2.1. OVERVIEW OF SICKLE CELL DISEASE (SCD)

Sickle-cell is regarded as the blood disorder and was asserted by Midence & Elander [1, 23]

RBCs are the major part of blood. Plasma is the vital liquid component of the blood on the other hand RBCs are also living part of the blood that are so tiny which can't be seen with naked eye. In blood the most abundant are RBCs beside them WBCs and platelets also found and there number ranges to millions. About 95% of RBCs consist of Haemoglobin (Hb) and most important function of them is to maintain the supply of O_2 to lungs and then to all body parts.

Haemoglobin is the predominant macromolecule in blood with a molecular weight of 64,500. According to world Health Organization report, normal level ranges between 120 and 130g/L for adults and abnormal concentrations lead to anaemia, Leukemia and Thalassemia [24].

Haemoglobin has a quaternary protein structure and consists of two pairs of different polypeptide chains designated as alpha (α) and the beta (β) chains. Each of the polypeptide chains is attached to a heme group which is a complex of iron and protoporphyrin. Thus, the haemoglobin molecule is a tetramer and binds four oxygen molecules [25]. Because of blood circulation, body organs and tissues get O_2 and this oxygen assists them for maintaining their activities and makes them alive.

Red blood cells that carry the normal haemoglobin are "doughnut" shaped. According to Tissot [26], disorder of haemoglobin synthesis such as sickle cell disease, alpha (α) and beta (β) Thalassemia's or haemoglobin variants – grouped under the term Haemoghbinopathies are frequently observed and up to 45% of new born from regions at risk present an abnormal haemoglobin. Individuals affected carry a mutated form of haemoglobin that distorts its normal shape and give them a sickled/cresent shape which becomes rigid/ stiffened [23, 24].

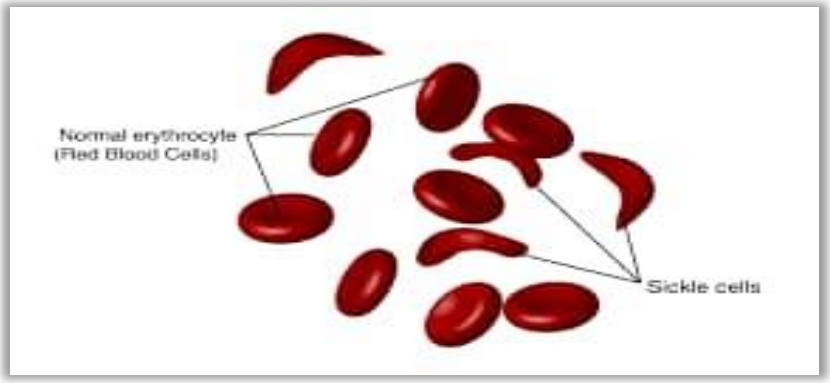

Figure 1: Normal Red Blood Cells (doughnut shaped) and Sickled Blood Cells[71]

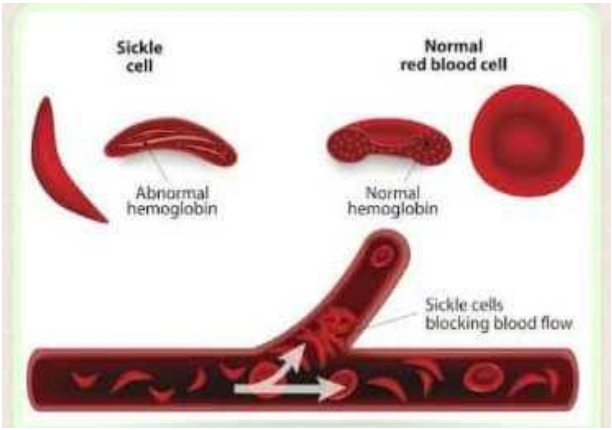

Figure 2: *SCA and Vasoocclusion of small capillaries [71].*

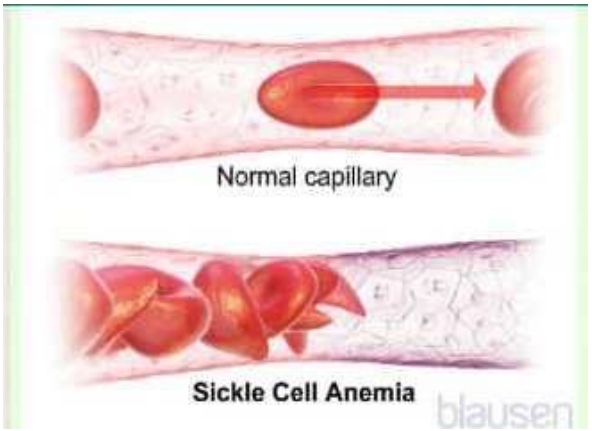

Figure 3: *Normal capillary flow and SCA [71]*

Sickle red blood cells cannot circulate freely and turn into stuck inside the capillaries, hence blocking the normal flow of blood [23]. The sickle cell disease/blood disorder is hereditary and not infectious.With two copies of sickle cell gene (HbSS or sickle cell anaemia), or the sickle cell mutation and another mutated beta-globin, for example sickle cell beta- thalassemia (HbS-beta-thal) or HbSC (mixed Haemoglobinopatries), the less soluble HbS can polymerize in deoxygenated regions of the circulation, resulting in red blood cell rigidity, RBC adhesion to the endothelium of blood vessel and haemolysis.

In addition to haemolytic anaemia, these events trigger the inflammation & coagulation cascade resulting vaso-occlusion [23, 27].

Thus, the clinical-disclosures are persistent haemolytic anaemia, frequent hurting episodes & continual organ-damage from vaso-occlusion. The affects of Sickle cell disorder appear in almost every organ or organ system of the body and results in renal failure. Hypertension, stroke and heart-diseases [28]. Most of the children die around the age of 5 years with severe form of SCA disease due to severe anaemia or infections.

SCD has key social & financial implications intended for the affected kid with family. Persistent sickle cell predicament affects the life of patient, particularly with respect to education, employment & psycho-social improvement. While most of the patients having SCD are usually well-adjusted, there is the risk of misery, low down self-esteem, communal seclusion, poor ancestral interactions & abandonment from usual daily-living. Awareness to psycho-social concerns are crucial for the psycho-social welfare & incorporation into the society of patients having SCD [5,1]

Presently, there is no cure for SCD. Therefore, choosing to become a parent comes with a lot of responsibility and people should take caution to gather information as possible about genetic diseases. However, individuals with sickle cell disease have specific issues and should seek the aid of genetic counsellors, coordinating testing, and discuss the probability for passing genetic disorder. This probability will be evidenced based according to the genotypes of both parents, allowing the potential parents to make a true informed decision prior to reproducing. [30].

2.2 GEOGRAPHICAL DISTRIBUTION OF SICKLE CELL VARIANTS

Sickle Cell Disease (SCD) is a serious blood condition caused by a single gene mutation that affects haemoglobin [31]. It has also been described as an inherited disorder of haemoglobin (Hb) in which the sickle haemoglobin (HbS) is present in association with normal haemoglobin HbA [32].

Worldwide the disorder due to single gene mutation & which is most common with 321,000 births per year is HbSS: globally the majority of such births happen in African sub-Sahara were approximately 300,000 babies are born annually with the disease [32,33]. Historically the homozygous HbSS disease, HbS beta-thalassemia, HbSC, HbSD and HbSE respectively are commonly included under the classification for SCD [34]. The geographical distribution of these HB variants differs and often parallels certain attributes such as climatic conditions and malaria endemicity. While HbSS and HbSC diseases are highly prevalent in areas of sub-Saharan Africa, particularly West Africa, the HbS-beta-thal, HbSD an HbSE are more common in parts of Middle East, Asia and Indian subcontinent [23, 35-37]. The prevalence of the carrier states for Sickle Cell Disease Hemoglobin AS the range was about 5% - 40% with the inhabitants in such prevalent areas & drives infections epidemiology [38]. The variant HbSS of (sickle cell anaemia) was found to be the most frequent among the Hb. It is the result of a single mutation in the haemoglobin gene and results from a GAG GTG nucleotide switch with a resultant substitution of valine for glutamic acid in the position 6 of the beta pleated globin chain, $\beta^{6Glu \rightarrow val.}$ [15]. This mutation creates a very significant functional defect in the Hb molecule. With two copies of the sickle gene (HbSS or sickle cell anaemia) or the sickle mutation and another mutated beta- globin gene, for example HbS-beta-thal (sickle cell-β^{O}-thalassemia), or HbSC (mixed Haemoglobinopathies), the Hb molecule becomes less soluble and in the event of low oxygen partial pressure or in deoxy-genated region of the movement it is increasingly polymerized, changing the structure of RBCs (Sickling), making them rigid or stiffened. This increases red cell interaction (adhesion) by vascular endo-thelium, white-cells (predominantly neutron-phils) & platelets. The devotion of Sickled Red-Cells to vascular endo-thelium & other cellular-events occurring in the blood triggers the irritation & coagulation cascade which leads to the vaso-occlusion/vascular-stasis, ischemia & RBCs sickling & eventual end-organ dysfunctioning.

In the order, HbSC, D & E results from $\beta^{6Glu \rightarrow lys}$, $\beta^{121Glu \rightarrow Gln}$, and $\beta^{26Glu \rightarrow lys}$ amino acid substitutions correspondingly [40].

In African Sub-Sahara, Benin, Cameroon, Senegal and in Nigeria particularly the SCD saddle is high which results the Burdon facilities of health as well as reduces the life quality and labour man losses.

2.3. NATURAL HISTORY AND NATURAL SELECTION

Sickle-cell syndrome was recognized in folk-medicines for centuries in various parts of Africa, but the emergence of Sickled RBCs was firstly reported in literature of medical about 1910AD as Herrick described it, the youngster Grenadian guy with periodic pain, anaemia & sickle-structured red-corpuscles inside the blood stream [41].

Historiographers consider that the mutations in DNA, that were responsible for 1st variants of sickle-cell gene, initially arise in diverse African-regions, including Benin Cameroon, Senegal and Central-African Republic [42]. Studies showed that the Trans – Atlantic slave's trade incorporated the sickle-cell genes to Americans & Caribbean-islands. African-slaves whom carried the sickle-cell attribute had β-globin specific genes. Pompa [40] expressed that the carriers that had sickle-cell traits, had hetero-zygote advantage and were being resistant to malarial infections.

Rewritten

In one study, Oludare [46] opined that the geographical distribution of the sickle cell trait is very similar to that of Malaria and this may explain why it has been maintained at such high prevalence levels in tropical Africa.

Figure 4: The geographic distribution of the five regions in which the sickle cell gene achieved high allelic frequency are superimposed on shading that identifies the old-world distribution of the sickle cell gene and the historic endemic malaria.

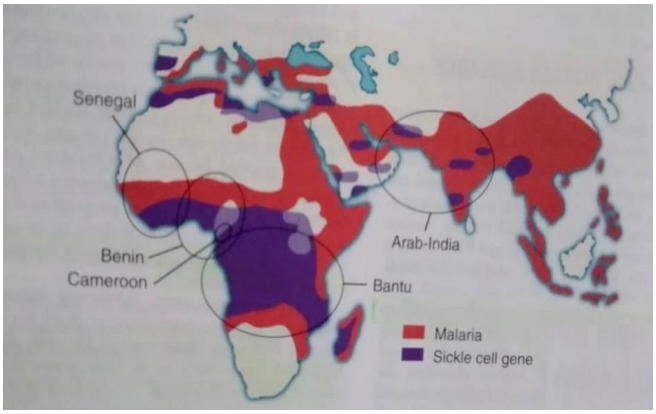

Figure 4: Geographic distribution of sickle cell gene and historical endemic malaria [8].

In regions with a high prevalence of the disease, falciparum malaria consistently affects young individuals and is the main factor leading to mortality in children having sickle-cell anemia (HbSS). Nevertheless, individuals with sickle cell trait had a reduced likelihood of experiencing high levels of parasitemia or severe malaria, particularly during early infancy. At the level of the (RBCs), this protection occurs following the first invasion of the parasite. One suggested mechanism connects the safeguarding of HbS to its instability, immunological state, & splenic-function. The infectivity of red blood cells with P. falciparum, in the folks having sickle-cell trait, causes a series of events. These include an increase in the denaturation of hemoglobin, the clustering of a particular membrane-protein called band-3, the hold of band-3 autoantibodies, the fastening of complement proteins, & an increased process of engulfing the early ring forms of the parasite by phagocytes. Thus, those with sickle cell characteristics may be safeguarded by a more rapid exclusion of parasitized RBCs by spleen, but those with homozygous HbS would no longer have this protection due to the development of functional asplenia. In the 1940s, Beet, a British Colonial Medical officer stationed in Zimbabwe, made the first observation that blood samples taken from malaria patients who had the sickle cell trait contained a lower number of Malaria parasites compared to blood samples taken from patients who did not possess the trait. In the 1950s, Anthony C. Allison formulated his own idea on the correlation between Malaria and the Sickle cell trait. Allison observed that those with the sickle cell trait in Zaire were less susceptible to malaria compared to those without the trait, based on her study of Beet and a physician at that period [44].

2.3.1. GENETICS AND PATTERN OF INHERITANCE OF SICKLE CELL DISEASE

Doss [53] stated that SCD is one of the prevalent hereditary haemoglobino-pathies worldwide. Hemoglobin HbA in adults is a diverse combination consisting of about 90% HbA, 2.5% HbA2, 3.5% HbA1c, and minor amounts of fetal hemoglobin [40]. Every typical hemoglobin molecule comprises IV polypeptide-chains, with 2 being β globin cuffs & 2 being α globin shackles. Sickle cell disease arises when the production of hemoglobin results in HbS rather than HbA. Sickle Cell Disease is a genetic condition that arises when Valine (Val) replaces Glutamic acid (Glu) at position 6 of the β globin chain [40].

Homozygous SCD, sometimes referred to as HbSS or Sickle Cell anemia, is a prevalent form of Sickle Cell beta gene inheritance occurring from both parents [54]. Sickle Cell haemoglobin SC is characterized by the presence of one S-globin chain and one β-globin chain, resulting in a relatively moderate characteristic. This gene is also present in the population of West Africans [55]. Furthermore, sickle Cell Sβº thalassemia and HbSS are categorized as more severe in comparison to other kinds

2.3.2. INHERITANCE PATTERNS

According to Osbourne [55], the following possibilities of inheriting the sickle cell disease or trait are plausible:

2.3.2.1. The sickle cell status of children when all the parents have the sickle cell gene

Sickle cell disease is a recessive genetic condition that manifests when an individual inherits two copies of the HbS gene, one from each parent. As shown in Table 1,

Table 1: Outcome of the interaction between two parents with sickle cell disease.

	Genotype of parents	Mother	
		S	S
Father	S	SS	SS
	S	SS	SS

2.3.2.2. The sickle status of children born to parents one with sickle cell disease and the other with sickle cell `trait.

all offspring of parents with two HbS genes will inherit the disease. When one parent has the sickle cell gene and the other has sickle cell trait (SCT), the probability of transmitting either gene to their offspring is equal, at 50%. In this scenario, each child has a 50% chance of inheriting sickle cell disease (SS) and a 50% chance of inheriting sickle cell trait (AS), as illustrated in Table 2.

Table 2: One Parent with Sickle cell trait and the other with Sickle Cell gene.

	Genotype of parents	Mother	
		S	S
Father	A	AS	AS
	S	SS	SS

2.3.2.3. The sickle cell status of offspring obtained when one parent has sickle cell disease (SS) and the other parent carries normal gene.

In this case, the HbA gene for normal Haemoglobin is carried by the normal parent and the gametes from this parent will carry this particular gene. Likewise, the sickle cell disease parent's gametes will carry the sickle cell Haemoglobin (HbS). All children born to these parents will carry the Sickle Cell Trait (carriers).

Table 3: One parent having Sickle Cell Disease and the other parent carrying normal genes.

	Genotype of parents	Mother	
		S	S
Father	A	AS	AS
	A	AS	AS

2.3.2.4. The sickle cell status of children obtained when all the parents have sickle cell trait.

When both of the parents carry sickle cell trait, the probability of their children inheriting the condition is 25%. There is an equal chance (50%) that their offspring will inherit the trait, and a 25% likelihood that they will have normal genes. In each pregnancy, there is a 50% chance that one parent will pass on the HbS gene and a 50% chance that the other parent will pass on the HbA gene. However, it's important to note that this doesn't guarantee a uniform distribution of genotypes among their children. Just as a couple may have all boys or all girls despite the 50% chance of each gender, it's possible for all children in a family to inherit the same genotype, whether SS, AS, or AA.

Table 4: The result of two parents with Sickle Cell Trait [55, 56].

	Genotype of parents	Wife	
		A	S
Husband	A	AA	AS
	S	AS	SS

2.3.2.5. The sickle cell status of children inherited from a relationship in which one parent has SCT and the other is normal.

In a case where one parent has SCT and the other is normal all children will display the SCT or have normal genes because half of the gametes of the parent that carries the SCT will carry HbS gene and the other half will carry the dominant gene HbA [55]. This combination will result in a 50/50 chance of producing a child with normal genes or inheriting the Sickle Cell traits. Again, take note this is just a chance game. Meaning you can have all children with normal genes as well as end up with Children all having the Sickle Cell trait as shown in table 5.

Table 5: One parent has SCT and the other is normal.

	Genotype of parents	Wife	
		A	S
Husband	A	AA	AS
	A	AA	AS

2.3.3 CLINICAL PRESENTATION OF SICKLE CELL DISEASE

The principal clinical symptoms of SCD include persistent hemolytic anemia, frequent bouts of discomfort, and chronic damage to various organs such as; spleen, skin, bones, brain, kidneys, lungs, & heart. The pattern of illness symptoms differs across the primary genotypes of HbSS, HbSC, and HbS-β-thalassemia, as well as within each genotype. The symptoms of the condition often appear during the first few months of birth, coinciding with a decrease in HbF levels and a rise in HbS levels [29,48].

One should be concerned about having SCD if he has:

- Unexplained pain in abdomen, chest, back, joints and muscles fatigue; the pain can be sudden in joints;
- Dizziness, fatigue, low oxygen in body and shortness of breath;
- Family history of sickle cell disease or trait;
- Inability to make concentrated urine or blood in urine;
- Hemolytic anaemia and anaemia that does not respond to iron supplements;
- Proof of SCA by Medical laboratory test;
- Painful erections in males;
- Yellowish skin colour and jaundice;
- Splenic sequestration (pooling) especially in children.

Specific age groups have a higher prevalence of certain problems. Individuals between the ages of 1 and 3 years experience an enlarged spleen, pooling of blood in the spleen, pneumonia & meningitis caused by Streptococcus pneumonia & further encapsulated-organisms, and hand-foot syndromes. In early childhood, they may have a stroke, acute chest syndrome, and osteonecrosis. In mid childhood, they experience pain crises, acute chest syndrome, and osteonecrosis. Between the ages of 12 and 20 years, they may have strokes, priapism, and pain crises. Between 20 and 30 years, they may develop renal insufficiency, pulmonary hypotension, disabling osteonecrosis, retinopathy, leg ulcers, and pain crises. At age 30 years and above, they may experience renal malfunctioning, congestive heart-failure, & pain criseses[29].

2.3.4. LIFE EXPECTANCY

Individuals with sickle cell disease (SCD) have a reduced life expectancy and a worse quality of life. The mortality rate from sudden cardiac death (SCD), especially among children under the age of five, is still unacceptably high. A significant amount of time is dedicated to being in the hospital due to the potential occurrence of a child's unexpected demise [50]. Life expectancy has declined, but there has been a significant improvement in the last 30 years, particularly in Western countries [29, 57, 58].

In 1973, Diggs documented that the average lifespan was 14.3 years. In 1994, Platt [59] documented that individuals with sickle cell anemia had a life expectancy of 42 years for males and 48 years for women. The ages for HbSC illness were 60 years and 68 years, respectively [41]. The enhanced lifespan may be attributed to advancements in overall medical treatment, such as the administration of preventive penicillin and vaccination against Streptococcus pneumonia [29, 48].

2.3.5. DIAGNOSIS

Sickle Cell Disease is suspected based on specific features in the Complete Blood Count (CBC) and peripheral blood film. The diagnosis is confirmed using hemoglobin electrophoresis [29, 60].

People with normal Hemoglobin usually have a haemoglobin level around 13.2-16.6 g/dl for men and 11.6-15/dl for women, with a normal hematocrit of 36.1% to 44.13% for Women and 40.7-50.3% for men. But people with SCD have a lower hemoglobin level, usually between 60-11g/dl and a low hematocrit, and a reticulocytosis (increase in number of immature RBCS) of approximately 3% to 15% [61].

Additional laboratory features of hemolysis are unconjugated hyperbilirubinaemia, elevated lactate dehydrogenase (LDH) and low haptoglobin levels. In the peripheral blood smear, they may be sickled RBCs, target cells and polychromasia indicative of reticulocytosis and Howell jolly bodies demonstrating hyposplenia. The RBCS are normochromic, normocytic and nucleated red cells are present. permanently sickled-cells (ISCs) are exclusively present around the marginal blood blotch of individuals with sickle cell disease, not in those with the sickle cell trait. In individuals with HbSS illness, there is a higher number of immature RBCs

(ISCs) and lower quantity of target cells. Additionally, there is a noticeable presence of smaller and paler red blood cells with a characteristic disc shape. Individuals with HbSS illness, especially those under the age of 10, have elevated levels of White Blood Cell Counts (WBCs) that exceed the typical range. In individuals with HbSS illness, the red blood cells are widely spaced, indicating a significant case of anemia [29, 62].

In resource limiting setting, the sodium metabisulfite test can be of diagnostic value. Adding sodium metabisulfite, a reducing agent to blood enhances deoxygenation of Hb and Sickling of HbS. The test does not distinguish Sickle Cell anemia from Sickle Cell trait or other HbS Syndrome because all cells sickle: however, sickling occurs more rapidly and with greater amount of HbS in the cells [62].

2.3.5.1. SOLUBILITY TESTS AND HEMOGLOBIN ELECTROPHORESIS

The solubility test findings (Sickledex) provide favorable outcomes for both Sickle Cell Disease (SCD) and Sickle Cell Trait. Every patient must have a conclusive diagnosis using Hb electrophoresis, a method that differentiates different types of hemoglobin based on their amino acid makeup. This is a conventional technique for distinguishing HbS from other variations [29, 62].

2.3.5.2. NEWBORN SCREENING

During the initial months of life; clinical symptoms of sickle cell disease (SCD), such as anemia, become apparent when the levels of HbS rise and HbF decline. Immature sickle cells (ISCs) are observable in the tangential blood-smear of infants suffering from sickle-cell anemia starting at 3rd month of life. By 4th month, somewhat brutal hemolytic-anemia becomes apparent [29].

2.3.5.3. PRENATAL DIAGNOSIS

A comprehensive study revealed that parents who were at a heightened risk of having a child with sickle cell disease (SCD) had a strong inclination towards prenatal diagnostics and expressed willingness to contemplate terminating a pregnancy if the baby was impacted by the condition. Chorionic villus sampling is used to acquire fetal DNA samples for prenatal diagnosis during the 8-10 week gestation period. PCR-based techniques are used to identify the sickle cell gene [29].

2.4. EPIDEMIOLOGY OF SICKLE CELL DISEASE

Sickle cell disease (SCD) mostly affects individuals of African descent. The World Health Organization has recently released a prevalence map of sickle cell disease (SCD) which indicates that approximately 20-15 million individuals worldwide have the HbSS genotype. Out of this population, around 12-15 million reside in Sub-Saharan Africa, 5-10 million in the Indian subcontinent, and approximately 3 million are distributed across Eastern Mediterranean states and Middle East populations, including countries such as Cameroon, Republic of Congo, Gabon, Ghana, and Nigeria. The prevalence of sickle cell trait (SCT) ranges from 20% to 30%, while in certain regions of Uganda, it can be as high as 45%. Approximately 2.3 to 3% of the population in these nations are afflicted with sickle cell disease [45, 46]. The prevalence of the sickle cell gene in Africa closely mirrors the occurrence of Malaria, namely caused by Plasmodium falciparum. The sickle cell gene is most prevalent in Africa's low-altitude tropical regions that see a substantial amount of yearly rainfall. Halden's 1949 study elegantly showcased the high occurrence of falciparum malaria and the distribution of HbS, β, and α-thalassemia. This study indicated that the heterozygous condition (HbAS) for sickle cell mutation may provide a survival benefit against severe malaria compared to the homozygous form. This phenomenon is referred to as balanced polymorphism [43]. The compound heterozygous HbSC is often seen in some regions of Africa, with a prevalence second only to the homozygous HbSS. However, it is characterized by a less severe illness pattern and mostly affects individuals of West African descent. The prevalence of HbS β-thal disease is considered to be low in Africa, with the majority of cases mostly found in Middle-East & Asia.

In South, East, & Central Africa, the occurrence of sickle cell disease (SCD) seems to closely resemble that of the Homozygous HbSS condition, with very few instances of other forms.

In 2006, the World Health Organisation (WHO) declared Nigeria to have the highest incidence of sickle cell anaemia (HbSS) in the world. The condition affects 2% of newborns and 0.05% of adults, with the lower frequency in adults being attributed to fatalities in early infancy [48, 49]. According to the global health watch, there are around 200,000 individuals in Africa who suffer from sickle cell anemia each year. It is worth mentioning that Nigeria alone has 150,000 sickle cell anemia children annually [50]. Remarkably, the globe has seen significant globalization and shifts in population dynamics due to migration, commerce, and the slave trade on an unparalleled scale in history. This has had a substantial effect on the epidemiology of SCD. The United States of America (USA), although not being considered endemic to Malaria, has a substantial population of individuals with Sickle Cell Disease (SCD). Approximately 100,000 individuals with SCD are believed to reside in the United States. The user's text is "[51]." The National Institutes of Health and Centers for Disease Control and Prevention [52] provide the following statistics:

- Sickle cell anaemia is the most common inherited disorder in the United States;
- More than 70,000 people in the United States have sickle cell;
- Sickle cell disease occurs 1 in every 500 African Americans;
- About 8% of African Americans are carriers of sickle cell disease;
- Two million people have sickle cell trait;
- Approximately 1 in 12 African Americans has sickle cell trait. In the United States, SCD accounts for less than 1% of all new cases of end-stage renal disease (ESRD).

2.4.1. COMPLICATIONS AND PROBLEMS ASSOCIATED WITH SICKLE CELL DISEASE

2.4.1.1. ACUTE PAIN

The hallmark of sickle cell disease is the sickle cell crisis which is an episode of pain. The hallmark of sickle cell disease is the sickle cell crisis which is an episode of pain. Acute pain is the first indication of illness in almost a quarter of patients and becomes the most common symptom after the age of 2. The primary reason why people with sickle cell disease seek medical assistance most often is due to the presence of pain [29, 48]. Marlowe and Chicella (2002) have consistently stated that Vasoocclusive events are the primary reason for hospitalization of sickle cell patients, resulting in yearly healthcare expenses of $475 million and hospitalization costs of $75,000 [29, 48].

The primary cause of pain is thought to be the blockage of blood vessels in the bone marrow, leading to tissue death known as bone infarction. This blockage occurs when the blood supply is cut off,

similar to what happens during a myocardial infarction. As a result of the tissue death, inflammatory substances are released, which then activate specialized sensory neurons called afferent nociceptors. These neurons are responsible for perceiving and transmitting pain signals. While the cause of most painful episodes in over half of sickle cell crises is unknown, the risk of experiencing a sickle cell crisis is increased by activities that increase the body's demand for oxygen, such as illness, physical stress, high altitudes, cold temperatures, dehydration, menstruation, and alcohol consumption [29, 48].

The sensation of pain is often experienced in the lumbar region, lower extremities, pelvic area, abdominal region, or thoracic region, sometimes manifesting in many sites. Bone pains are prevalent because to the direct harm caused by blood blockage (vasooclusion) to the bone(s) and the fact that bone marrow is responsible for the production of red blood cells. Acute Chest Syndrome (ACS) is a condition characterized by the deprivation of oxygen to the lung tissues during a crisis. The experience may be very agonizing, perilous, and perhaps fatal. It is the primary cause of sickness among individuals with sickle cell disease (SCD) and is the prevailing state at the time of death. ACS is often associated with pulmonary infections, which may be caused by viruses, bacteria, or fungus.

2.4.1.2. INFECTIOUS DISEASES

Infections are prevalent and may lead to significant complications. Parvovirus B19 infection is a frequent cause of temporary interruptions in the production of red blood cells (aplastic crisis) in individuals with sickle cell disease (SCD). This condition is characterized by sudden decreases in hemoglobin levels, reticulocyte count, and red blood cell precursors in the bone marrow. It is important to note that there is no obligatory increase in lactate dehydrogenase (LDH) levels, particularly in children. However, the presence of protective antibodies in adults significantly reduces the likelihood of parvovirus causing aplasia in this age group. According to reports, Acute chest syndrome, glomerulonephritis, hepatic sequestration, pulmonary fat embolism, and bone marrow necrosis have all been shown as outcomes of this [29, 48]. Infections of the urinary tract and pyelonephritis are more common in people with sickle cell disease (SCD) than in the general population. Patients with UTIs are at risk for septicemia due to the prevalence of Escherichia coli, the most common uropathogen. ACS bouts often occur due to bacterial infections, particularly with Mycoplasma pneumonia.The microorganisms mentioned include Streptococcus pneumoniae, Chlamydia pneumoniae, Legionella spp, and Haemophilus influenzae. Antibiotics are administered to individuals with pneumonia or ACS to address infections caused by these organisms [29]. The primary etiology of osteomyelitis is often attributed to Salmonella spp.

2.4.1.3. ERECTILE DYSFUNCTION

Approximately 30% of individuals with sickle cell disease may have persistent erectile dysfunction, either partially or completely. Approximately 40% of men, including youngsters with sickle cell disease, have priapism. Priapism is a medical disease characterized by an enduring and painful erection that is not caused by sexual arousal and does not subside with sexual activity. This occurs as a result of the obstruction of blood flow in the penile arteries by sickled cells, and it may persist for a duration ranging from few hours to several days.

Impotence is the major issue that arises from priapism, however individuals who have had many bouts of priapism may nevertheless report satisfactory erections and continue to engage in sexual activity. The objective of therapy in this case is to alleviate priapism and preserve erectile function. While this anecdotal data supports the use of hydralazine in treating acute priapism, it is crucial to prioritize avoiding the recurrence of priapism as part of the overall care approach. Strategies include the delivery of HU, chronic transfusion, the use of the antiandrogen bicalutamide, and self-administration of the α-adrenergic drug etilefrine orally or by intravenous injection during episodes lasting over 1 hour, among other approaches. Hydroxyurea is known for its ability to increase the amount of HbF, reduce the number of white blood cells, enhance the hydration and lifespan of red blood cells, and reduce inflammation and adhesiveness of red blood cells [29, 41, 48].

2.4.1.4. GALL BLADDER DISEASE

Cholecystitis, also known as gall bladder disease, is often seen in individuals with sickle cell disease and is closely correlated with the rate of hemolysis. Approximately 30% of children diagnosed with sickle cell disease experience the presence of gallstones, whereas by the age of 30, this percentage increases to 70% among patients. Typically, gallstones remain asymptomatic for many years. Patients experiencing symptoms may exhibit postprandial satiety, have discomfort in the right hypochondrium, or manifest nausea and vomiting. Acute episodes may be mistaken for a sickle cell liver crisis, but often, ultrasonography is utilized to definitively diagnose gallstones [29, 48].

2.4.1.5. LIVER DISEASE

More than 50% of individuals with sickle cell disease have hepatomegaly, whereas acute liver injury affects up to 10% of patients admitted to the hospital [48]. Acute hepatic cell crisis is characterized by the presence of a swollen liver that is painful to touch, along with a worsening of yellowing of the skin and eyes (jaundice) and an elevated body temperature (fever). The probable cause is ischemia of the hepatocytes. The levels of AST and bilirubin are increased. This illness often resolves with the provision of supportive treatment, but it has the potential to advance to liver failure and result in a catastrophic outcome. Consequently, it is crucial to constantly evaluate patients and commence exchange transfusion if they exhibit indications of advancing liver disease, such as a rise in AST levels. Acute hepatic sequestration is characterized by a sudden increase in liver size and a significant decrease in hemoglobin content. The primary reason for this is the accumulation of sickled red blood cells in the liver. The approach to treatment involves the administration of supportive care and transfusions.

People with sickle cell illness are more likely to get viral hepatitis since they need transfusions so often. Perhaps it is the most important element in the progression of cirrhosis. One successful therapy for this issue is liver transplantation. Screening methods for donated blood have decreased the hazard of administering interferon-ribavirin to SCD patients for the treatment of hepatitis C and B [29, 48].

2.4.1.6. BONE COMPLICATIONS

Some children with SCD have hyperplasia of blood cells in the bone marrow, leading to aberrant bone growth, which manifests as elongated limbs or deformed heads. Vaso-occlusion, which impedes oxygen supply to the bone, may also lead to osteoporosis

and discomfort. Children experiencing sickling in their hands and feet may develop a distressing condition known as hand and foot syndrome. The intense bone pain that often arises around the age of 2 is frequently the first indication of sickle cell disease (SCD). This condition of dactylitis often cures on its own and may be managed with enough water and pain relief medication. Avascular necrosis of the hip, a disorder that affects about 50% of adult sickle cell patients, develops when the bone tissue dies due to a lack of oxygen supply. Ultimately, adult patients may need surgical intervention to excise deteriorated and necrotic osseous tissue. Patients with severe instances may need joint replacement surgery [29,48].

2.4.1.7. DERMATOLOGIC COMPLICATIONS

Leg sores and ulcers are significant contributors to illness and disease burden in individuals with sickle cell disease due to their high occurrence, long-lasting nature, and limited response to treatment.

Spontaneous or trauma-induced wounds may become infected, with the most frequent bacteria involved being Staphylococcus aureus, Pseudomonas spp, Streptococci spp, or Bacteroides spp [29]. Sickle cell disease (SCD) may also result in vascular damage in the eye, leading to retinal scarring and detachment that can ultimately result in blindness (48). The retina is more susceptible to vasoocclusion, and it is recommended that patients with SCD get yearly retinal examinations as part of their standard healthcare maintenance [29].

2.4.1.8. OBSTETRIC AND GYNECOLOGICAL ISSUES

Women with SCD are more prone to have gynecologic issues such as delayed onset of menstruation, painful menstruation, ovarian-cysts, pelvic-infection, & fibro-cystic sickness of breast. Women having sickle-cell disease (SCD) whom get pregnant has an increased chance of developing difficulties for both themselves and their child, however these risks are not significant enough to prevent them from continuing with the pregnancy. The fetal problems of pregnancy, mostly caused by compromised placental blood flow, include a higher occurrence of unprompted premature abortion, intra-uterine developmental retardation, low birth weight infants, and fetal mortality. The symptoms of SCD tend to worsen during pregnancy, with painful episodes becoming more frequent. The individual is experiencing severe anemia due to deficiencies in iron or folate. This is accompanied by an exacerbation of the normal anemia that occurs throughout pregnancy. Additionally, there is an increased susceptibility to infections such as urinary tract infections, pneumonia, and endometritis. The individual is also at risk for developing preeclampsia and potential mortality. These findings are supported by [29,48]. Nevertheless, by implementing meticulous prenatal care and surveillance, it is possible to prevent severe complications [48].

2.4.1.9. BIRTH CONTROL

It is advisable to explore using modern intrauterine devices that do not contain estrogen. Administering depot injections of medroxyprogesterone (Depo-Provera) every 3 months seems to have a low risk of causing strokes [29]. Patients with sickle cell disease (SCD) are at risk of experiencing bone loss, which is an important factor to address due to their tendency to have skeletal issues. Oral-contraceptives contain minimal amounts of estrogen may be considered without any conclusive evidence of an elevated risk of stroke seen so far, while the patient's total risk of stroke should still be taken into account. It is recommended that women who are sexually active get regular pelvic examinations and receive guidance on birth control methods [29].

2.4.1.10. PULMONARY COMPLICATIONS AND HYPERTENSION

SCD is mostly responsible for causing death in persons with pulmonary illness. Both acute and chronic pulmonary issues are often seen. Pneumonia and acute chest syndrome are the most severe issues, but the main long-term effect is raised blood pressure in the pulmonary circulation, also known as pulmonary hypertension. This condition is caused by increased resistance to blood flow in the arteries that supply the lungs. Pulmonary hypertension may develop in older children and adult individuals who have sickle cell disease (SCD). Approximately 60% of patients have persistent pulmonary issues, such as pulmonary hypertension [29, 48].

2.4.1.11. STROKE

Stroke is the second leading cause of death among sickle cell disease patients over the age of three (03) years. Those with SCD had a much greater relative risk of stroke, ranging from 200 to 400 times, compared to those without SCD [29]. Strokes often occur as a result of obstructions in the blood arteries responsible for transporting oxygen to the brain. This issue might arise abruptly and unexpectedly. Research has also shown that around 8-10% of individuals with sickle cell disease get strokes, usually around the age of 6. Research has also shown that by the age of 14, around 8% of individuals with sickle cell disease (SCD) have had a stroke, and by the age of 20, this number increases to 11%. Among Cameroonian patients, sickle cell anemia (SCA) is the primary cause of stroke in children, accounting for 31.4% of cases [29, 48, 63-65]. Transcranial Doppler Ultrasonography (TCD) and magnetic Resonance imaging (MRI) are diagnostic tools for strokes. The TCD assesses stroke risk levels by monitoring the average blood velocity [49, 66]. Individuals with sickle cell disease (SCD) are also more susceptible to experiencing strokes due to an aneurysm, which is a weakened blood vessel wall that may burst and result in bleeding. Multiple aneurysms are often seen in individuals with sickle cell disease; however, their location generally renders them unsuitable for surgical intervention [48].

2.4.1.12. KIDNEY DISEASE/RENAL COMPLICATIONS

Renal injury is a common issue in sickle cell anemia due to the harsh conditions in the renal medulla, such as low oxygen levels, increased acidity, and high osmolarity. These conditions encourage the formation of HbS polymers and lead to negative alterations in blood viscosity. Indeed, this phenomenon also happens in individuals with the sickle cell trait. The renal cortex shows increased blood flow, which is thought to be responsible for glomerular dysfunction and the presence of albumin in the urine [41, 48, 67]. Hypertension, proteinuria, hematuria, worsening anemia, and nephritic syndrome are reliable indicators of the development to renal failure. These clinical signs should be closely monitored since they provide valuable information, especially because blood creatinine levels may be deceptive. Individuals diagnosed with sickle cell anemia have an elevated proximal excretion of creatinine [29]. Urinary issues are quite prevalent, especially nocturnal enuresis. Patients may have hematuria, which is the presence of blood in the urine. However, this symptom is often moderate, painless, and disappears without causing any harm. Kidney failure is a significant risk to elderly people and is

responsible for 10-15% of fatalities in individuals with sickle cell disease [48].

2.4.1.13. ANAEMIA

Chronic hemolytic Anemia is a significant hallmark of the sickle cell disease commonly called sickle cell anemia. Anemia that is primarily due to increased red blood cell destruction is called Hemolytic Anemia [62]. RBC life span in sickle cell anemia averages about 15days but with marked inter individual variability from 7 to 30 days. In HbSC disease, the average is about 30 days [41]. Because of the short life of the Sickle Red Blood Cells, the body is unable to replace red blood cells as quickly as they are destroyed. This causes a particular form of anaemia called Hemolytic anemia. Most patients with sickle cell disease have hemoglobin levels of about 5g/dl, much lower than healthy persons. Chronic anaemia reduces oxygen levels and increases the demand on the heart to pump more oxygen bearing blood through the body. Eventually, this can cause the heart to become dangerously enlarged, with an increased risk for heart attack and heart failure [48].

Progressive anemia from renal endocrine deficiency or a decrease in bone marrow function from vasooclusion is associated with early death [29].

2.4.2. MANAGEMENT OF COMPLICATIONS IN SCD AND TREATMENT

Despite advancements in the administration and care of sickle cell disease, as well as bone marrow transplantation, it still remains a significant source of mortality and morbidity in Nigeria, Cameroon, Benin, and other regions of Africa. Nevertheless, the treatment required for SCD is of a prolonged duration due to its chronic nature, resulting in economic and psychological consequences for both patients and their families [67]. The two main objectives of treatment are to modify the illness and avoid crises, complications, chronic organ damage, and early death. Additionally, therapy aims to provide compassionate, timely, effective, and safe relief for acute crises, including pain episodes. Thus, the primary focus of outpatient clinic treatment is to implement strategies aimed at preventing pain crises, alleviating symptoms, avoiding organ problems, and enhancing survival rates [29].

Without universal health insurance, families bear the responsibility of caring for chronic illnesses like sickle cell disease (SCD). When multiple family members are affected, SCD imposes a significant financial burden on the family, potentially leading to ongoing conflicts and financial strain [49, 56].

Presently, individuals diagnosed with sickle cell disease get medical attention in secondary and tertiary hospitals, as well as some private healthcare facilities. However, this treatment lacks a well-defined framework, a functional system for referring patients, and the integration of targeted therapies for varying levels of care. The recommended approach for controlling sickle cell disease at the community level in Nigeria is a comprehensive set of measures designed to decrease mortality and the occurrence of complications in people with SCD. The contents of the bundle include:

- Raising Public awareness;
- Newborn screening;
- Screening for SCD at primary health care centres;
- Registry of patients with SCD for prospective follow-up;
- Prophylaxis for infection, pneumococcal vaccines, oral penicillin use of insecticide treated bed nets and antimalarials;
- Health maintenance and comprehensive care;
- Genetic counseling of individuals with abnormal haemoglobin; AS, SS, SC;
- Nutrition;
- Education of patients and care givers about sickle cell disease, including what to do in acute conditions before coming to the hospital;
- Optimal hydration by teaching the patients to drink enough fluids to make their urine clear and whitish without yellow colour [49, 68].

In Consonance with Nnodu [49], Adebowale [48] opined that if one has been diagnosed with sickle cell disease, one can decrease the frequency of pain crisis by following some simple guidelines:

- Maintain good nutrition including supplement of folic acid, zinc and vitamin E;
- Drink plenty of fluids especially during hot weather exercise or when travelling;
- Plain water and fruit juices are best choices;
- Use of over the counter medication, warm baths, heating pads, fluids and bed rest at the first indication of onset of a pain episode;
- Use acupuncture, between feedback and relaxation to reduce stress of the disease.

Healthcare workers and academics still face significant challenges in managing and preventing bouts of pain. While a definitive treatment for sickle cell disease has not yet been discovered, there is potential for enhancing the care provided to people with this condition. Prabhakar [69] observed a deficiency in the comprehensive comprehension of the pathophysiology of SCD. Additionally, he found that the major emphasis in SCD clinical care is on the treatment of problems rather than the prevention of difficulties. Complications may be prevented by using infection prevention, which includes many measures.

2.4.2.2. MALARIA PREVENTION

Malaria is linked to poor health and fatalities in regions with a high prevalence of malaria, such as Nigeria, Cameroon, Benin, and others. While there is no consensus on the use of a particular drug for preventing malaria in children living in areas where malaria is common, the guidelines from the federal ministry of Health in Nigeria recommend several measures. These include educating people about health, controlling the environment, spraying insecticides indoors, distributing bed nets treated with insecticides, and using a drug called proguanil for prevention. The recommended dosage of proguanil varies depending on the age of the child, ranging from 25mg for children under one year old to 100-200mg for children over three years old.

2.4.2.2. VACCINATION AND ORAL PENICILLIN V PROPHYLAXIS

It is important to vaccinate children against S. pneumoniae, Haemophilus influenzae, hepatitis B, and influenza, among other diseases. Administering vaccines and using Penicillin prophylaxis may effectively decrease the likelihood of severe pneumococcal infections. Penicillin prophylaxis starts at the age of 2 years. The recommended preventive penicillin dosage for children under the age of 5 is 125mg of penicillin V administered orally twice a day until the age of 2-3 years, and then increased to 250mg for children

older than three years. Children up to the age of 16 and patients with a history of pneumococcal sepsis should get this preventive treatment. Patients with a history of pneumococcal sepsis should continue taking penicillin prophylaxis permanently. For individuals with a penicillin allergy, it is recommended to provide Erythromycin at the same dosage.

The recommended vaccination schedule advises administering the heptavalent pneumococcal conjugated vaccine at 2 months of age, followed by two more doses spaced 6-8 weeks apart. A booster dose should be given at 12 months. Immunization should be provided in accordance with the national protocol outlined in references [29, 49].The timetable for immunizing children with sickle cell disease in Nigeria, as per the official standards, shown in the table.

Table 6: schedule of immunization of children with sickle cell disease, according to the national guidelines

Immunizations	Period
BCG	At birth
OPVO	At birth
OPV1	6 weeks
OPV2	10 weeks
OPV3	14 weeks
Hepatitis B1	At birth
Hepatitis B2	6 weeks
Hepatitis B3	14 weeks
Haemophilus influenza	3, 6,10, 14 weeks
DPT	6,10, 14 weeks
Pneumococcal conjugate 13 valent vaccine	From 6 weeks, monthly x 3 doses and a booster at 12 months, followed by pneumococcal polysaccharide vaccine 23 valent from 2 years with booster dose 3 years later
Measles	9 months
Yellow fever	9 months

BCG = Bacille Calmette Guerin Vacline ; OPV = Oral Penicillin V ; DPT = Diphtheria, Pertussis and tetanus vaccine

2.4.2.3 VITAMIN OR NUTRITIONAL SUPPLEMENTATION AND HYDRATION

Research has shown that individuals with sickle cell disease (SCD) in sub-Saharan Africa have fewer episodes of crises, fever, and hospital visits when their diet and hydration are properly addressed [68].

Persistent breakdown of red blood cells leads to higher use of folic acid reserves. It is recommended that all patients be given folic acid supplementation at a dosage of 2.5 - 5mg per day, taken orally. This should be provided as part of routine therapy [29,49]. Patients with SCD might also have vitamin B12 insufficiency. Substitutes for folate may conceal and perhaps worsen a shortage in vitamin B12. Emerging research suggests that individuals with sickle cell disease often have shortages in many minerals and vitamins, such as Zinc, vitamin C, E, D, A, Calcium, and Acetylcysteine, among others.A recent prospective experiment shown that Zinc supplementation resulted in weight increase in infants with Sickle Cell Disease (SCD). Hence, it may be beneficial to take a multivitamin without iron on a regular basis. Furthermore, patients with a significant deficit of vitamin D may consider further vitamin D replacement treatment.

Children with Sickle Cell Disease in the communities should be advised to consume water in the morning, mid-morning, and mid-afternoon under supervision, and often throughout the day to ensure that their urine, which is typically yellow, turns clear. Individuals diagnosed with sickle cell disease have impaired ability to concentrate urine, making them susceptible to dehydration if they do not consume sufficient fluids (recommended at a rate of 60ml per kilogram of body weight per 24 hours in adults). Intravenous hydration is used when the patient is not consuming an adequate amount of fluids orally. It is preferable to maintain a urine specific gravity of 1.01 by daily testing. In addition, it is recommended to provide them with a meal rich in protein, as well as a nutrition package that includes fruits and vegetables [29].

2.4.2.4. STROKE MANAGEMENT

Stroke is a very destructive consequence of SCD. The occurrence is 300-fold greater in those with HbSS compared to those with HbAA [49]. Sickle cells have the ability to obstruct the circulation of blood to a specific region of the brain. A stroke occurs when there is a significant interruption in the quantity of blood flow to the brain, leading to a reduction in the supply of oxygen [70]. Complications of sickle cell disease (SCD) may arise suddenly and manifest as recurrent seizures, muscle weakness or numbness in the extremities, and unexpected difficulty with speech and loss of consciousness. These signs and symptoms should prompt quick medical treatment for anybody caring for a patient with SCD [71]. Transcranial Doppler Ultrasound Scan (TCD USS) and magnetic resonance imaging (MRI) are both diagnostic tools for identifying strokes. The TCD assesses the risk levels by quantifying the Mean blood velocity [49, 66]. A chronic blood transfusion program has been discovered to decrease the TCD USS velocities in children who are at a high risk of stroke [29, 72, 73]. This intervention is costly and beyond the capabilities of nations without well-established blood transfusion programs, relying on replacement or commercial donors for their blood supply. Consequently, individuals with Transcranial Doppler (TCD) Ultrasonography (USS) Velocities over 200cm/s will have blood transfusion in order to decrease the level of hemoglobin S to less than 30%. Subsequently, they will be prescribed hydroxyurea therapy. Individuals with a Velocity ranging from 170 to 199cm/s will have a repeated Transcranial Doppler Ultrasound (TCD USS) examination after three months. If they are identified as being at high risk, they will be provided with the appropriate therapy as previously outlined.

Chronic transfusion may be worsened by the occurrence of alloimmunization, which involves the production of antibodies against blood type K, C, E, S, F, Y, and JK antigens. In order to reduce the occurrence of this, if feasible, blood from the HbAA group, which has been carefully matched for these antigens, will be used. Transfusion in Sickle Cell Disease (SCD) is aimed at

decreasing the level of HbS to less than 30%. Therefore, HbAA is the recommended choice for transfusion, rather than HbAS [49, 74]. Yogen S (29) suggests that the most effective method for avoiding alloimmunization is to decrease the number of leukocytes and do restricted phenotypic matching for all patients, including ABO, C, D, E, and Kell antigens. Additionally, extended phenotype matching should be performed for patients with any antibodies. The transmission of human immunodeficiency virus, hepatitis B and C, and human T-cell Leukemia/lymphoma virus-1 has decreased due to enhanced screening of stored units. However, it still poses a concern. Furthermore, the implementation of improved screening systems may be complemented by the use of leukocyte-depleted RBC transfusions, which can effectively mitigate this risk.

2.4.3 TREATMENT

Several medicines may serve in the therapy process. Hydroxyurea (HU) is a widely used chemotherapeutic agent that inhibits the growth of bone marrow cells and is licensed by the FDA for the treatment of Sickle Cell Disease Patients. It was approved in 1998 [75]. Hydroxyurea has been used as a chemotherapeutic drug for cancer treatment for a considerable period of time. Individuals who have both sickle cell disease (SCD) and Hereditary persistence of fetal hemoglobin (HPFH) have 70% HbS in their red blood cells (RBCs), but do not experience anemia or other related symptoms. Research indicates that the equal distribution of fetal hemoglobin (HbF) in red blood cells (RBCs) disrupts the formation of HbS polymers, enhances its ability to dissolve, and inhibits the process of sickling. These results led to the suggestion that using drugs to stimulate the synthesis of HbF might be advantageous for people with sickle cell disease [29]. Hydroxyurea is now used to reduce the process of sickle cell hemoglobin (HbS) Polymerization and erythrocyte sickling by promoting the formation of fetal hemoglobin (HbF). This, in turn, has many advantageous effects on the structure, content, and function of red blood cells (RBCs). This decreases the need for blood transfusion and the probability of organ impairment [75]. This medication has effectively decreased the frequency of painful episodes and the likelihood of recurring complications associated with sickle cell disease (SCD). Physicians advise that persons who often suffer painful episodes or have a history of acute chest syndrome, symptomatic anemia, and other Vasoocclusive crises may consider hydroxyurea treatment [76]. In a randomized placebo-controlled experiment, adults with hemoglobin SS who were being treated with hydroxyurea saw a significant decrease in vasoocclusive crises and the occurrence of acute chest syndrome due to hydroxyurea medication. According to Moore [29], Hydroxyurea has been identified as a cost-effective therapy for sickle cell anemia. Another medicine, L-glutamine (Endari), in the form of oral powder, is intended to aid in the prevention of crises and, thus, reduce the likelihood of hospitalization. Crizanlizumab-tmea (adakveo) prevents the aggregation of blood cells and the subsequent obstruction of narrow blood arteries, a condition that may cause both discomfort and organ damage [77].

2.4.3.1. OVER THE COUNTER MEDICINES

Across the nation, several medications include nonsteroidal anti-inflammatory drugs (NSAIDs), acetaminophen (Tylenol), Ibuprofen (Advil or Motrin), as well as opioids such as codeine, morphine, and oxycodone, are used for the management of sickle cell pain. Non-opioid medications may enhance the pain-relieving effects of opioids when used together. Acetaminophen is a commonly prescribed medication for managing sickle cell disease. Acetaminophen is considered a very safe analgesic and it works by blocking the production of prostaglandins. However, NSAIDs are contraindicated for people with gastritis, peptic ulcer, and renal failure. Opioids are often used analgesics for managing moderate to severe acute pain caused by pain episodes [78].

2.4.3.2. TRANSFUSION THERAPY

Transfusions are an additional approach used to address issues associated with SCD and aid in the prevention of strokes and acute chest syndrome. Transfusions are advised for individuals with sickle cell disease (SCD) who have acute chest syndrome, heart failure, severe anemia (in youngsters), enlarged spleens, multi-organ failure, splenic sequestration aplastic crisis, and are prone to strokes [79]. A clinical trial assessing the effectiveness of transfusion found that it decreases the likelihood of recurring strokes in children with SCD. This research also shown that 50% of children with sickle cell disease (SCD), who had experienced strokes and had not had transfusions, would get strokes within a period of three years, in contrast to just 10% of those with SCD who had received transfusions.

2.4.3.3. BONE MARROW TRANSPLANT

Another therapeutic intervention administered to individuals with sickle cell anemia is Bone Marrow Transplantation (BMT). The phenomenon is being reported more often, yet it remains contentious despite promising outcomes. Opponents of this treatment for sickle cell anemia express worry about the acute and long-term consequences, morbidity, and expense in comparison to standard therapy [48].Currently, transplantation (specifically allergenic transplantation) is only recommended for people with sickle cell disease who have a severe enough condition to warrant the associated risk [80].

2.4.3.4. EXPERIMENTAL TREATMENT

Gene therapy is an innovative medical approach used to treat individuals suffering from sickle cell anemia. Gene therapy is considered superior than blood transfusion by researchers due to its capacity to provide a long-lasting remedy. Gene therapy necessitates a delivery mechanism capable of transporting a functional gene to cells harboring faulty genes. Researchers have selected genetically altered viruses as a very effective method of transportation. Target cells are infused with regular genes to guarantee their proper functioning [42]. Gene therapy research is still at its nascent stage, and several experts are of the opinion that this approach has the potential to serve as the definitive treatment for SCA.

2.4.4 PREVENTION OF DISEASE OCCURRENCE

Sickle cell disease (SCD) is considered a public health priority by the World Health Organization (WHO) because of its significant prevalence [5]. Although hydroxyurea and L. glutamine are commonly used and effective medications for managing sickle cell disease (SCD), there are still significant challenges and unmet requirements in treating this condition. Many patients continue to have poor clinical outcomes in both the short and long term [81]. Despite advancements in its treatment, such as enhanced care and bone marrow transplant, the disease continues to result in premature mortality and morbidity among children in Cameroon, Nigeria, and other regions of Africa [65]. Stem cell transplantation is the most promising treatment option for achieving a cure, but it

is seldom performed due to the considerable dangers and exorbitant expenses associated with it. Due to the absence of a pharmacotherapeutic remedy for SCD, the management approach mostly focuses on palliative care. This involves mainly maintaining good health and addressing both acute and chronic problems. Therefore, it is advisable to avoid the birth of a child who would be impacted by Sickle Cell Disease (SCD), since this might ultimately result in the complete elimination of the condition. According to Dr. Artemos Francis, a medical practitioner located in Kaduna, Nigeria, the first measure to address the problems caused by Sickle Cell Disease (SCD) is to focus on preventing its onset. According to his argument, sickle cell can only manifest when two individuals who possess the sickle cell trait procreate. It is crucial to enhance counseling for "intending couples" to undergo testing in order to know their sickle cell status before making informed choices about reproduction [48]. Adeyemo [82] states that strategies for avoiding the birth of children with haemoglobinopathies (SCD) including prenuptial (premarital) screening and genetic counseling, Utero–therapy by stem cell transplantation, and prenatal and preconception diagnosis [57, 4, 82]. Nevertheless, experts unanimously agree that the most effective strategy for preventing Sudden Cardiac Death (SCD) is to identify carriers via genetic counseling and testing. Genetic counseling is economically efficient in low-income areas with a high prevalence of the condition [67].

Implementing premarital screening and genetic counseling programs has the potential to decrease the incidence of infants being born with sickle cell disease (SCD) and other haemoglobinopathies [82, 83]. The institution of marriage is the fundamental basis of any civilization. Premarital counseling is a kind of guidance provided to young people or couples-to-be in order to prevent potential issues that may arise in married life, such as a troubled marriage or even divorce, particularly in cases involving the loss of a child. Mounting evidence indicates that genetic premarital counseling and testing, especially for sickle cell disease, may effectively decrease the occurrence of hereditary Haemoglobin diseases. This is primarily achieved by identifying and providing counseling to couples who are at a high risk of passing on these illnesses. Crucially, prenatal counseling has been shown to have a significant benefit compared to neonatal screening. Prenatal counseling focuses on primary prevention, while neonatal screening deals with secondary or tertiary prevention, often after the harm has already occurred [57]. A research conducted by Memish and Saeedi found that there was a consistent decline in the prevalence of haemoglobinopathies, from 32.9 to 9.0 per 1000 individuals evaluated, throughout a period of 6 years of premarital screening [57].

The effectiveness of preconception counseling and testing for haemoglobinopathies relies on the target population's awareness of sickle cell disease (SCD), their attitude towards genetic counseling and screening, and their comprehension of the implications of having a child afflicted by the disease [57, 64, 82]. A cross-sectional study conducted at King Khalid University examined the knowledge, attitude, and practice of students regarding pre-marital counseling (PMC). The study found that educational level significantly influences the viewpoints and outcomes of PMC, regardless of the cultural beliefs prevalent in the region [84]. 96.5% of students expressed their voluntary intention to attend PMC after an education and awareness campaign, whereas 72.1% said that PMC should be obligatory.

Authors have suggested further strategies to prevent and eliminate illness, such as prenatal diagnosis and the termination of pregnancies afflicted by the condition [57]. During the embryonic stages, the fetus undergoes testing to see whether it has any genetic disorders. whether it is confirmed to be a carrier, the pregnancy is terminated during the first three months.

Nevertheless, the lady is compelled to have several pregnancies, but only a small number of them are tested to confirm that they do not contain any genetic disorders and are thus permitted to survive. In reality, this is not only costly but also poses a risk to the life of the lady in issue.

The Ministry of Health in Bahrain introduced the premarital counseling program for sickle cell disease (SCD) at all health clinics in 1993. In 2004, a royal decree was issued, making it mandatory for all couples intending to marry to undergo premarital screening and counseling. In 2007, a program for screening newborns for hematological abnormalities was created [9]. Regrettably, in Cameroon, the national control programme for Sickle Cell Anemia (SCA) has not been put into action. There has been no implementation of a screening programme for newborns or retrospective screening in the public sector. Additionally, Cameroon does not currently have a specialized center for providing lifelong medical care and monitoring for individuals with SCA. In addition to preventative genetic services, such as prenatal diagnosis of SCA, they have not yet been integrated into the national health systems. Prenatal genetic diagnosis is a preventative method that allows parents to screen babies that are at risk and make choices about terminating affected pregnancies [85]. In Cameroon, prenuptial testing is offered, however there is a low participation rate because to insufficient health promotion efforts to raise awareness. While the importance of disease management via preventative methods such as public education, genetic counseling, and screening has not been adequately highlighted, it is a more cost-effective technique, particularly in settings with limited resources [5]. The effectiveness of a disease prevention program is often contingent upon the level of knowledge and attitudes of both healthcare professionals and the people impacted by the condition [85]. Nelson Mandela, the South African anti-apartheid revolutionary leader, said that education is the most potent tool one can use to effect global transformation.

If premarital counseling is effective in preventing high-risk couples from marrying, it can prevent families from facing the lifelong challenge of individuals living with sickle cell disease. This disease goes beyond the clinical complications and painful crises, causing significant conflicts and social consequences for patients and their relatives (9). The results of premarital counseling are thought to be affected by the counselors' approach in providing the couple with information on the dangers, complications, and future treatment of the illness. The potential for counselors to use an insufficient approach may result in the couple being denied their rights to receive complete information, which may have a negative impact on their ability to make informed decisions and exercise their autonomy. In Nigeria, premarital genetic counseling for sickle cell disease (SCD) is optional, although many religious organizations often require genetic testing before couples may be united [46, 86].

MATERIALS AND METHODS

3.1 STUDY DESIGN

This study was a community based cross-sectional observation study.

3.2 STUDY SITE

- **Tubah Council**

Tubah Council is located in Mezam Division in the North West Region of Cameroon. It is situated between several Councils; Belo Council in Boyo Division, Bafut Council, Bamenda III Council, Balikumbat Councils and Ndop Councils in Ngoketungia Division and Santa Council. It lies strategically along the ring road, crisscrossing two Divisions – Boyo and Ngoketungia. Tubah Subdivision covers an area of 450 square kilometers with an estimated population of 65,200 inhabitants drawn from the four main villages; Kedjom Keku, Kedjom Ketinguh, Bambui (including Finge and Baforkum) and Bambili that make up the municipal Council area.

Agriculture is the main economic activity in the area. Among other functions carried out by the Council is the licensing of marriages, drawing up of birth certificates and death certificates, assistance to health services and educational services in the area (Annex 5)

- **Bamenda I Council**

The Bamenda I Council came into existence following Decree No. 2007/117 of 24th April 2007. It has a surface area of 110 square kilometres with a population of 62,000 inhabitants.

Bamendankwe is the only village that makes up the Council area. Bamendankwe is a large village with about 24 quarters

The Bamendankwe people are mainly of the Tikar origin from the locality of Menda near Bankim in the now Adamawa Region though for either socio-economic or administrative purposes, many people have migrated to the area, mainly the Bamilekes. The settlement in this area took place between 1902-1903 and coincided with the arrival of the Germans between 1906-1908.

Agriculture is the main economic activity of the people, with more women involved in the art. Other activities include: cattle rearing, market gardening, Art and Craft and Tourism.

The Bamenda I Council was created to carter for the administrative, security, educational and health needs of the ever-growing population. (Annex 4)

- **Bamenda II Council**

The council was created following presidential decree Nº 2007/115 of 13 April 2007 and Nº 2007/117 of 24 April 2007. It covers a total surface area of 1,482km^2 with a projected population of 261,285 persons in 2019 and a population density of approximately 176.3 persons per kilometers square. The high density is as a result of concentration of administrative, health and educational and socio-economic institution in the Council centre. The main ethnic group is the Ngemba tribe. It has 4 main villages, Mankon, Chomba, Nsongwa and Mbatu. Agriculture, mining, (quarry exploitation), forestry, and craftsmanship make up the economic life wire of the inhabitants. (Annex 3)

- **Bamenda III**

The Bamenda III Council was created by Presidential Decree No. 2007/115 of 13 April 2007 and 2007/117 of 24 April 2007. It is located in Bamenda Subdivision. Bounded to the East by Tubah municipality, to the South by Bamenda I, to the West by Bamenda II and to the North by Bafut municipality. It is the gateway to and from the Divisions of Boyo, Ngoketungia, Bui and Donga/Mantung. It covers a total surface area of 22.9km^2 hectares and has an estimated population of 105,244 inhabitants (according to projection by CAMGIS in Minimum Urban Local Development Scheme (SMAUL) Feb. 2008. It has two main Clans, Nkwen and Ndzah, with more than 25 communities.

The Bamenda III Council was created with the aim to bring administration and governance nearer the people, improve access to healthcare/staffing existing healthcare units and also improving the road infrastructure/network to ease access to healthcare units and markets.

Perfect social infrastructure and provide education to the people. Since then, it has been carrying out these activities to improve the welfare of the population not without difficulties though (Annex 1)

- **Bamenda City Council**

Bamenda is the largest and most influential administrative and commercial centre in the North West Region. Creation of the Bamenda City Council stemmed from establishment of Bamenda as a regional administrative centre by the colonial leaders.

The Bamenda City Council covers the Divisional Headquarters of Mezam and the Sub divisional Headquarters of Bamenda, I, Bamenda II and Bamenda III according to the administrative system in Cameroon.

Bamenda City Council has a surface area of 1076 kilometres square with a population estimated at 322,889 inhabitants according to the 2005 population general census for Cameroon. Out of this population, 269,530 inhabitants constitute the urban population while 53,359 is suburban.

The important potential of Bamenda City emanated from the fact that it is the first City in the North West Region and the commercial heart bit of North West. Also, it is the gateway City to Western and South West Region of Cameroon and a gateway for Trans-African road to Central African Republic and Federal Republic of Nigeria.

The City is fast growing and extending into the rural settlements of Mankon, Nkwen, Mbatu, Bamendankwe, Ndzah, Chomba and Nsongwa areas (Annex 2)

3.3. STUDY DURATION

The study was carried out for a period of 04 months, from March to June 2022.

3.4. STUDY POPULATION

The population targeted by this study was drawn from Tubah, Bamenda I, II, III and City Councils. In Tubah Council, 13 workers voluntarily accepted to participate by signing the inform consent out of 58 employed workers. In Bamenda I, 11 out of 43 workers enrolled. Meanwhile in Bamenda II and III, 08 and 11 out of 84 and 43 workers respectively participated. The City Council registered the highest number of respondents: 17 out of 253 workers, making a total of 60 respondents.

3.5. SELECTION CRITERIA

3.5.1. INCLUSION CRITERIA

All council workers who accepted voluntarily to participate by signing the inform consent were included in the study.

3.5.2. EXCLUSION CRITERIA

All council workers unable to sign the written informed consent and who returned incompletely filled questionnaires were excluded from the study.

3.6. SAMPLING TECHNIQUE

A convenient sampling technique was employed to get Council Workers and Council staff who gave their consent to participate in the study. The investigator approached the respondents at their workplace at convenience and after explaining the purpose of the study, assigned questionnaires to respondents who became interested in the study.

3.7. SAMPLE SIZE

A total of 59 respondents who voluntarily accepted to participate in the study by signing the informed consent were enrolled for the study.

3.8. DATA COLLECTION INSTRUMENT

A semi-structured questionnaire was used to gain an in-depth understanding of the knowledge, attitude and practices of respondents in the Tubah, Bamenda I, II, III and City Councils with respect to premarital counseling and screening for SCD.

3.9. DATA COLLECTION PROCEDURE

After obtaining ethical clearance, respondents who met the inclusion criteria were advised on the proper way to fill the informed consent and then proceeded unto the questionnaire itself. The survey assessed the level of knowledge on SCD premarital counseling and testing using a questionnaire comprising of 18 knowledge questions (Annex 1)

3.9.1. Score of Knowledge

Each of the knowledge questions carried 01 mark. Respondents were said to have:

- excellent knowledge when they scored ≥14 out of 18;
- very good knowledge ≥ 11 out of 18;
- good knowledge ≥9 out of 18;
- and poor knowledge when they scored strictly <9 out of 18.

3.9.2. Score of attitudes

Attitude of respondents was assessed using the aforementioned questionnaire comprising 13 questions carrying a mark. (Annex1). Respondents were considered to have:

- positive attitude if they scored > 9 out of 13 questions;
- and negative attitude if they scored < 9 questions out of 13.

3.9.3. Score of Practices

Assessment of respondent's practices towards sickle cell disease premarital counseling and testing was done on Section C of the aforementioned questionnaire carrying 11 questions.

Respondents who had 8 out of 11 questions correctly were said to have adopted adequate practice and below 8 correct questions was taken for inadequate practice. Positive score indicates positive attitude and practices while negative score indicates poor attitude and practices.

3.10. DATA MANAGEMENT AND ANALYSIS

All the completed questionnaires were received, verified and validated by the investigator. A descriptive analysis was done using statistical package for Social Sciences (SPSS) version 21. Categorical variables were presented as frequency, percentages and charts. Chi square test was used to test for association between sociodemographic variables and knowledge, attitude and practices of respondents towards SCD premarital counselling and testing.

The survey used a *P-value* of <0.05 as a cut of point for statistical significance.

3.11. ETHICAL CONSIDERATION

Ethical clearance was sought and obtained from the Institutional Review Board of the University of Bamenda. Administrative authorization was obtained from the Regional Delegate of Public Health for the North West Region and the Mayors of the Tubah, Bamenda I, II, III and City Council involved in the survey (see appendix).

RESULTS

4.1. SOCIODEMOGRAPHIC CHARACTERISTICS OF RESPONDENTS

Table 7 presents the socio-demographic characteristics of respondents. Out of a total of 59 respondents, the majority (64.4%) were 38 years and above. These were followed by those between 29 to 38 years (20.3%). Females were more represented (67.8%) than males (32.2%). The majority of the respondents were Christians (98.3%). Only 1 respondent was Muslim (1.7%). With respect to the level of education, majority of the respondents (39%) had advanced level. These were followed closely by graduates (35.6%). Those who endedat secondary level of education came with 20.3%. According to the marital status of the respondents, 61% were married, 37.3% not married and 1.7% still in union. Regarding positions held by respondents in the various councils, 33.9% worked in the civil status registry. Technical staff represented 22%, administrators 15.3% and sanitary officers 11.9%. 16.9% of workers played other functions in the councils.

Table 7: Socio-demographic characteristics of respondents (N=59)

Variables	Characteristic	Frequency (N=59)	Percentage (%)
Age range (years)	19-28	9	15.3
	29-38	12	20.3
	>38	38	64.4
Gender	Male	19	32.2
	Female	40	67.8
Religion	Christianity	58	98.3
	Islam	1	1.7

Educational level	Primary	3	5.1
	Secondary	12	20.3
	Graduate	21	35.6
	Advanced level	23	39.0
Marital status	Married	36	61.0
	In union	1	1.7
	Not married	22	37.3
Status in Council (position)	Civil status	20	33.9
	Sanitation	7	11.9
	Administration	9	15.3
	Technical staff	13	22.0
	Others	10	16.9

4.2. KNOWLEDGE OF COUNCIL WORKERS ON SICKLE CELL DISEASE PREMARITAL COUNSELLING.

4.2.1: Overall Knowledge of study participants.

Majority of the council workers had poor knowledge (61.9%), followed by those who had very good knowledge (19.0%), good knowledge (16.7%), and then excellent knowledge (2.4%),

(**Fig. 5).**

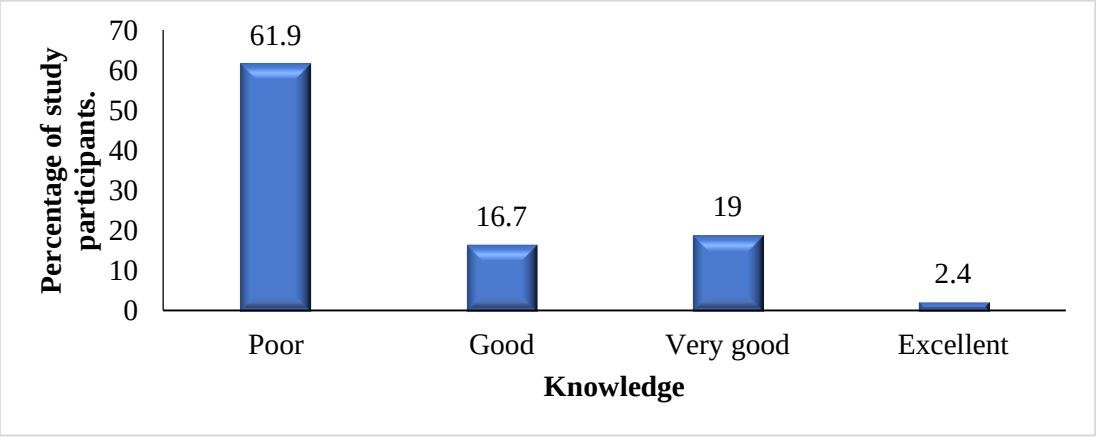

Figure 5: Distribution of study participants with regards to knowledge of Sickle cell disease and sickle cell premarital counseling.

4.2.2 Representation of study participants with regards to source of information on sickle cell disease.

Majority of the participants (45.3%) heard about sickle disease from their friends and relatives. 24.5% heard from health professionals, 17% from school, 7.5% from TV/Radio, 3.8% from the internet and 1.9% from conversation in the quarter. (**Fig. 6).**

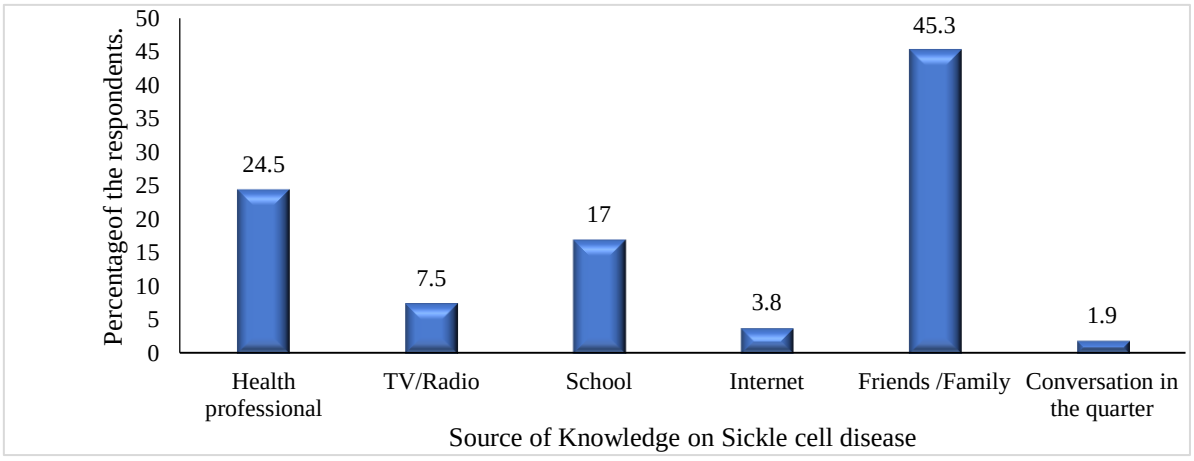

Figure 6: Distribution of study participant with regards to source of knowledge of Sickle cell disease.

4.2.3) Association between knowledge and Socio-demographic factors

As shown on table 8, there was a significant association between level of education and participant's knowledge on sickle cell disease (X^2 = 17.5, P= 0.04). All those with primary level of education had poor knowledge (100%). Majority of those with secondary education had good knowledge (57.1%) while majority of those who had advance level rather had poor knowledge (73.3%). As regards graduates, more than half (52.9%) had poor knowledge. For sex, more females (71.4%) had poor knowledge than males (38.5%), these results were however not statistically significant (X^2 = 6.0;P= 0.11). All those in the younger age group of 19 – 28years had poor knowledge (100%), the same as thosefrom 29 – 38 years (71.4%). Meanwhile for those greater than 38 years, a greater number had poor knowledge (48.1%). These results were however not statistically significant (X^2 = 7.5, P= 0.28).

Table 8: Association between knowledge and socio-demographic factors

VARIABLES	Poor	Good	Very good	Excellent	X^2	*P*-value
	N (%)	N (%)	N (%)	N (%)		
Sex						
Male	5(38.5)	3(23.1)	5(38.5)	0(0.0)		
Female	20(71.4)	4(14.3)	3(10.7)	1(3.6)	6.0	0.11
Age group/years						
19 - 28	8(100.0)	0(0.0)	0(0.0)	0(0.0)		
29 - 38	5(71.4)	1(14.3)	1(14.3)	0(0.0)	7.5	0.28
>38	13(48.1)	6(22.2)	7(25.9)	1(3.7)		
Level of Education						
Primary	2(100.0)	0(0.0)	0(0.0)	0(0.0)		
Secondary	3(42.9)	4(57.1)	0(0.0)	0(0.0)		
Advance level	11(73.3)	0(0.0)	3(20.0)	1(6.7)	17.5	**0.04***
Graduate	9(52.9)	3(17.6)	5(29.4)	0(0.0)		
Marital status						
Married	10(43.5)	6(26.1)	6(26.1)	1(4.3)		
In union	1(100.0)	0(0.0)	0(0.0)	0(0.0)	8.81	0.18
Not married	15(83.3)	1(5.6)	2(11.1)	0(0.0)		
Religion						
Christianity	26(63.4)	7(17.1)	7(17.1)	1(2.4)		
Islam	0(0.0)	0(0.0)	1(100.0)	0(0.0)	4.35	0.23
Status in Council						
Civil status	8(72.7)	1(9.1)	2(18.2)	0(0.0)		
Sanitation	4(80.0)	0(0.0)	1(20.0)	0(0.0)		
Administrator	3(42.9)	1(14.3)	2(28.6)	1(12.5)	11.38	0.5
Technical staff	3(37.5)	4(50.0)	1(12.5)	0(0.0)		
Others	4(57.1)	1(14.3)	2(28.6)	0(0.0)		

N;Frequency, %; percentage of study participants, X^2; Chi-square value

4.2.4: Participants' knowledge on PMCSCD in the different councils

Participants in all the councils had poor knowledge. 35.7% of those in city council had good knowledge. Also, City council came first for very good knowledge with 21.4%. For excellent knowledge Bamenda III came first with 10%. These findings were not statistically significant (X^2= 14.2, P = 0.29) (**Table 9).**

Table 9: Knowledge of respondents on PMCSCD in the different councils

Council	**Poor** N (%)	**Good** N (%)	**Very good** N (%)	**Excellent** N (%)	X^2	P-value
Bamenda I	6(85.7)	1(14.3)	0(0.0)	0(0.0)		
Bamenda II	4(66.7)	0(0.0)	2(33.3)	0(0.0)		
Bamenda III	6(60.0)	1(10.0)	2(20.0)	1(10.0)	14.2	0.29
City council	6(42.9)	5(35.7)	3(21.4)	0(0.0)		
Tubah council	4(80.0)	0(0.0)	1(20.0)	0(0.0)		

N; Frequency, %; percentage of study participants, X^2; Chi-square value.

4.3. ATTITUDE OF COUNCIL WORKERS ON SICKE CELL DISEASE PREMARITAL COUNSELLING AND SCREENING.

4.3.1: Type of attitudes

More than half of the council workers had good attitude towards sickle cell disease and sickle cell disease premarital counselling (54.2%), **(Fig.7).**

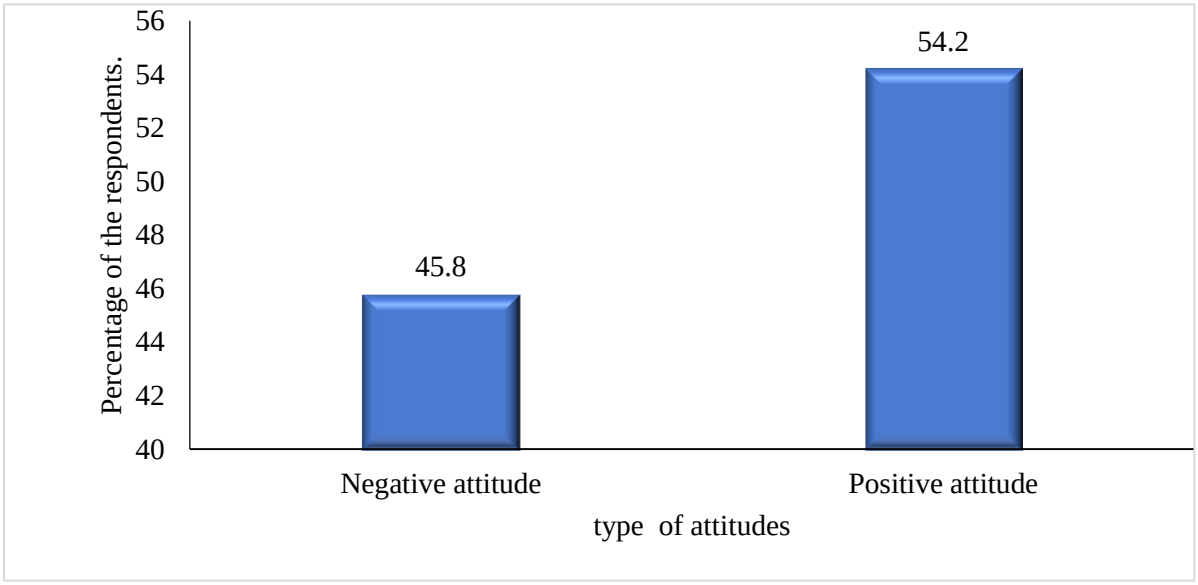

Figure 7: Attitude of council workers towards sickle cell disease premarital counselling

4.3.2 Association between attitude and socio-demographic factors

More males (73.7%) than females (43.6%) had positive attitude towards sickle cell disease and sickle cell premarital counselling. These results were statistically significant (X^2= 4.6, P = 0.03). As regards age group, all those from 19 to 28 years had negative attitude (100%). Majority (66.7%) of those 29 – 38 years had negative attitude while majority (73.7%) of those >38 years had positive attitude. These results were statistically significant (X^2= 18.6, P<0.001). For marital status, the married (71.4%) had positive attitude than the unmarried (31.8%). These findings were statistically significant (X^2= 9.8;P = 0.007). There was no significant relationship between attitude and level of education as well as between attitude and religion.

Table 10: Association between attitude and socio-demographic factors

VARIABLES	Negative N (%)	Positive N (%)	Total N (%)	X^2	*P*-value
Sex					
Male	19(100)	0(0.0)	19(100%)	0.51	0.48
Female	37(97.4)	1(2.6)	38(100%)		
Age group(years)					
19 – 28	9(100)	0(0.0)	9(100%)		
29 – 38	10(90.9)	1(9.1)	11(100%)	4.35	0.11
>38	38(100)	0(0.0)	38(100%)		
Level of education					
Primary	2(100)	0(0.0)	2(100%)		
Secondary	11(91.7)	1(8.3)	12(100%)		
Advance level	22(100)	0(0.0)	22(100%)	3.81	0.28
Graduate	21(100)	0(0.0)	21(100%)		
Marital status					
Married	35(100)	0(0.0)	35(100%)		
In union	1(100)	0(0.0)	1(100%)	1.6	0.44
Not married	21(95.5)	1(4.5)	22(100%)		
Religion					
Christianity	56(98.2)	1(1.8)	57(100%)		
Islam	1(100)	0(0.0)	1(100%)	0.02	0.89
Status in Council					
Civil status	16(94.1)	1(5.9)	17(100%)		
Sanitation	6(100)	0(0.0)	6(100%)		
Administrator	8(100)	0(0.0)	8(100%)		
Technical staff	12(100)	0(0.0)	12(100%)	2.16	0.7
Others	10(100)	0(0.0)	10(100%)		

4.3.3 Participants' attitude towards PMCSCD in the different councils

As shown on table 11, participants in Bamenda I and Bamenda II councils were the least in positive attitudewith 44.4% of their workers showing positive attitude towards sickle cell disease and sickle cell premarital counselling. Participants in Bamenda III council showed the highest positive attitude (66.7%), followed by those from Tubah council (63.6%) then those from city council (52.9%). However, these results were not statistically significant (X^2= 1.83, *P*= 0.77).

Table 11: Participants' Attitude in the different councils

Council	Negative N (%)	Positive N (%)	Total	X^2	*P*-value
Bamenda I	5(55.6)	4(44.4)	9		
Bamenda II	5(55.6)	4(44.4)	9		
Bamenda III	4(33.3)	8(66.7)	12	1.83	0.77
City council	8(47.1)	9(52.9)	17		
Tubah council	4(36.4)	7(63.6)	11		

N; Frequency, %; percentage of study participants, X^2; Chi-square value

4.4. PRACTICE OF COUNCIL WORKERS ON SCD PREMARITAL COUNSELING.

4.4.1: Type of practices

Up to 96.6% of the council workers had poor practice of sickle cell disease premarital counselling (Fig. 8).

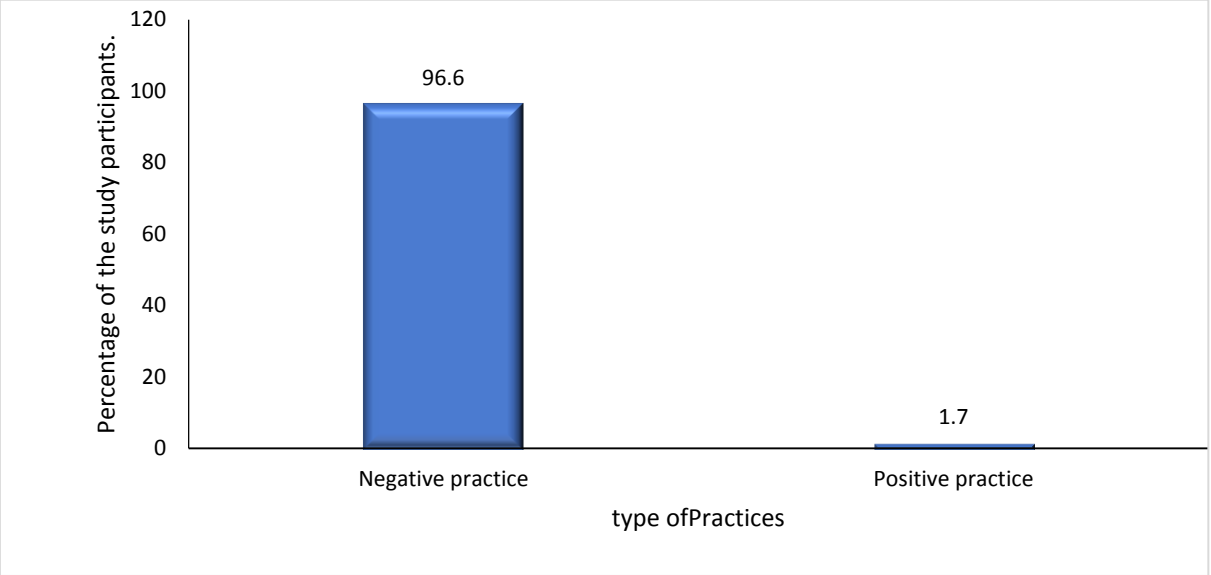

Figure 8: Participants' practices of sickle cell disease premarital counselling.

4.4.2 Association between respondent's Practices of PMCSCD and Socio-demographic factors

In this survey, more males (100%) have negative practice than females (97.4%). However, these findings were not statistically significant (X^2 = 0.51, P=0.48). All those aged 19 to 28 years and >38years had negative practice (100%), while 90.9% of those 29 to 38 years also had negative practice. These findings were also not statistically significant(X^2 = 4.35, P=0.11). Equally, there was no significant association between practice and level of education, marital status, religion and status of council worker(Table 12).

Table 12: Association between Practice and Socio-demographic factors

VARIABLES	Negative N (%)	Positive N (%)	Total N (%)	X^2	P-value
Sex					
Male	19(100)	0(0.0)	19(100%)	0.51	0.48
Female	37(97.4)	1(2.6)	38(100%)		
Age group(years)					
19 – 28	9(100)	0(0.0)	9(100%)		
29 – 38	10(90.9)	1(9.1)	11(100%)	4.35	0.11
>38	38(100)	0(0.0)	38(100%)		
Level of education					
Primary	2(100)	0(0.0)	2(100%)		
Secondary	11(91.7)	1(8.3)	12(100%)		
Advance level	22(100)	0(0.0)	22(100%)	3.81	0.28
Graduate	21(100)	0(0.0)	21(100%)		
Marital status					
Married	35(100)	0(0.0)	35(100%)		
In union	1(100)	0(0.0)	1(100%)	1.6	0.44

Not married	21(95.5)	1(4.5)	22(100%)		
Religion					
Christianity	56(98.2)	1(1.8)	57(100%)		
Islam	1(100)	0(0.0)	1(100%)	0.02	0.89
Status in Council					
Civil status	16(94.1)	1(5.9)	17(100%)		
Sanitation	6(100)	0(0.0)	6(100%)		
Administrator	8(100)	0(0.0)	8(100%)		
Technical staff	12(100)	0(0.0)	12(100%)	2.16	0.7
Others	10(100)	0(0.0)	10(100%)		

N; Frequency, %;Participants,$X^{2;}$ Chi-square value

4.4.3 Participants' Practice of PMCSCD in the different councils

But for city council with 5.9% of positive practice, participants from all other councils had negative practice (100%). The results were not statistically significant (X^2 = 2.45, P=0.65). Table 13

Table 13: Participants' Practice in the different councils

Council	Negative (%)	Positive (%)	Total	X^2	p-value
Bamenda I	9(100)	0(0.0)	9		
Bamenda II	9(100)	0(0.0)	9		
Bamenda III	12(100)	0(0.0)	12	2.45	0.65
City council	16(94.1)	5(5.9)	17		
Tubah council	11(100)	0(0.0)	11		

DISCUSSION, CONCLUSION, LIMITATIONS AND RECOMMENDATIONS

5.1. DISCUSSION

This survey presents one of the SCD community findings about the knowledge, attitudes and practices of Council workers towards premarital sickle cell disease counseling and testing in Cameroon. According to data analysis, more females contributed to the study than males.

A similar trend was observed in other studies by Tusuubira *et al.* [13] and AL-ghubishi *et al* [60], where females contributed a higher percentage (87.9%) of all responses than males. This discrepancy could have happened during distribution of questionnaires whereby more females received questionnaires than males. It may also be that most men were away for work or due to men's negative attitude as highlighted by Tusuubira *et al.* [13]. Another factor could be that more females were willing to fill the questionnaire than males. The majority of the sample population was in the age category of 38 years and above, followed by 29-38 years. With respect to educational level in this study, majority 39%, (23) reported to have had advance level, closely followed by graduates (35.6%) who reported to be graduates. Nearly all respondents were Christians (98.3%) with only one respondent that was Muslim (1.7%). As for marital status, the survey found more married respondents (61%) than singles (37%). This result is inconsistent with a study by Al-ghubishi *et al.* [60], who reported more singles (89.5%) as against 7.9% married. The discrepancy here could be associated to the component of population under study, whereby respondents in this survey were wage earners (workers), as against high school attendants in the former. Out of the sociodemographic characteristics of interest, level of education was found to be significantly associated with knowledge (X^2=17.5; P=0.04). This means that education is vital in the campaign to reduce the prevalence of SCD.

In the knowledge assessment section, majority of the respondents in this study had poor knowledge score (61.9%) on SCD and SCD premarital counseling. This result is not the same as results obtained in a survey in Yaba Nigeria by Oludare *et al.*[46] where 80% of respondents had proper knowledge of SCD and SCD premarital counseling but concurs with results obtained in rural

areas of Nigeria (17.8%) and Sudan (25.7%) with exceptionally low knowledge scores on SCD [87]. The poor knowledge level amongst council workers of the Tubah , Bamenda I,II,III and City council could be due to poor sensitization and awareness campaigns and is a wakeup call for implementation of SCD premarital counseling programs in these councils to increase awareness.As for the source of information in this study, family and friends appeared to be the most important source (45.3%) followed by health professionals (24.5%), schools (17%), TV/Radio (7.5%), Internet (3.8%) and conversation in the quarter (1.9%) . This is the same with results obtained in a study by Tusuubira *et al*.[13] and wonkam *et al.* [65] but not true in the case of Ngwengi *et al*.[5] who demonstrated that school ranked highest as a source of SCD knowledge in studies conducted in Ghana and Nigeria. This result trend is to be expected since family is considered the primary source of knowledge for most people. One participant is quoted as saying "My parents always cautioned us from marrying from a certain family, because always heard somebody hospitalized in that family" [13]. The small proportion that obtained information from health care professionals and schools in this study, suggests that there is limited efforts in health care settings and schools to inform the public on SCDEven though council workers of the city council came first with very good knowledge (21.4%), followed by Bamenda III with excellent knowledge (10%), these findings were not statistically significant(X^2=14.2; P=0.29).

In the attitude assessment section of our survey, more than half the council workers had good attitude towards sickle cell disease premarital counseling (54.2%).Even though a majority of respondents reported not knowing their sickle cell status, 90% showed willingness to know. More males than females had positive attitudes towards sickle cell disease premarital counseling. These results where statistically significant (X^2 = 4.6, P=0.03). More males showed positive attitudes because men are the folk that decide in issues of marriage and so tend to seek for more knowledge. Participants in the age group 19 – 28 years had negative attitudes (100%). The negative attitudes in this young age group is similar to that reported in a study by Oludare *et al*. [46]. This could be because this age group falls within the adolescent. They are adventurous and want to go the way of love (infatuated) and do not bother the consequences or lean towards religious believes on miracles and destiny. This might also imply that more sensitization is required for this group of persons. Majority of respondents in the age group 29 – 38 years had negative attitudes too whilethose > 38 years had positive attitudes. The negative attitudes in the age bracket 29-38 years could relate to individuals who are becoming more desperate to get married or get engaged. Meanwhile the positive attitude in the age group>38 years towards SCD and SCD premarital counseling isprobably due to exposure from past life experiences. For marital status, the married showed a positive attitude as against the unmarried (X^2 = 9.8, P=0.007). Other studies by Al-ghubushi *et al* .[60] have reported a better knowledge and attitude in married people compared to the unmarried our attitude results towards SCD premarital (54.2%) were similar to results obtained in a previous study made in Saudi Arabia with 42% of the participants in Riyadh and 29% in Abha supporting implementation of compulsory premarital genetic counseling law [87]. However, the variation in attitude shown by some participants in this study is consistent with studies By Al-ghubushi *et al*.[60] in which Saudis showed different attitudes towards sickle cell disease premarital counselingin a study carried in Jazan. Participants in Bamenda I and Bamenda II were the least in positive attitudes with 44.4% of the workers agreeing to sickle cell disease premarital counseling. Council workers in Bamenda III showed the highest positive attitudes (66.7%), followed by those from Tubah council (63.6%) and then city council 52.9%.

Out of all the three aspects observed in our studies, the participants performed the worst in practice. Up to 96.6% of the council workers had poor practice with regards to sickle cell disease premarital counseling. This is consistent with the study by Al-ghubushi *et al*. [60] , where only 19.1% of participants show good practice, in our survey results no socio-demographic variable was found to be significantly associated with practice. With the exception of the city council with 5.9% positive practice, participants from all other councils had 100% negative practice.The only reason that could be advanced in this study for the poor practice is lack of knowledge. Lack of knowledge is intricately linked to poor practice because in practical life you can never give what you do not have. Poor practice amongst participants may also indicate the lack of law regulation and governmental monitoring regarding premarital counseling for SCD.

5.2. CONCLUSION

The results from this survey show that;

- Council workers have poor knowledge of sickle cell disease and SCD premarital counseling.
- Attitude of council workers towards SCD premarital counseling was good.
- Practice of premarital counseling for SCD among study participants remained poor**.**

5.3. LIMITATIONS OF STUDY

- The small sample size which affects our ability to generalize the findings.
- Limited resources did not allow the investigator to assess the Knowledge, attitude and practices of Council workers in other places in Cameroon which can provide a nation-wide understanding of the current state of knowledge, Attitudeand practice and the need for national awareness program.

5.4. RECOMMENDATIONS

From the results of the survey, this study therefore recommends as follows:

- Local government Councils should pull attention to potential areas for intervention and improvement in knowledge of man power resources especially true in relation to SCD counselors so that this knowledge can be translated to intending couples to foster better attitudes and practices in sickle cell awareness, prevention and management.
- The Minister of Public Health should include SCD in existing health education programs both at community and health Centre settings/levels. Education should target various organizations such as schools, religious, social and health bodies but should start with the parents who will intend educate their children not to allow love to blind fool them into future problems whereby they will not enjoy their marriages as a result of having children that will make them go in and out of hospital at any time.

- The Ministers of Secondary and Higher Education should emphasize SCD studies in the curriculum of students especially at the tertiary levels to increase awareness because it is at this stage that many young stars meet their spouses. Proper education will bring about improved knowledge on the disease transmission and symptomatology and will lead to positive attitude and good practices.

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