KNOWLEDGE AND PERCEPTION OF SICKLE CELL DISEASE AND PRE MARITAL HAEMOGLOBIN GENOTYPE SCREENING AMONG NATIONAL YOUTH SERVICE CORPS MEMBERS IN OYO STATE

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DEDICATION

This work is dedicated to my late Sister, Mrs. Hope Amata for her love and support towards my success while she was alive.

ABSTRACT

Sickle Cell disease has become recognized as a World Wide problem affecting millions of people. Despite the common occurrence of the disease, public awareness is low with the condition often attributed to cultural beliefs such as "ogbanje" and "abiku" (the children born to taunt their parents by dying young).

Beliefs change when younger, educated generations such as National Youth Service Corps Members (NYSC) pass on their knowledge to the community where they serve as teachers, health and extension workers. This research sought to find out whether NYSC Members' knowledge of sickle cell disease and aspects of prevention, like haemoglobin genotype screening, is adequate.

This is an exploratory descriptive survey of 291 Corps members in Oyo State. The study population consisted of all 840 NYSC members posted to Oyo State in October 1990. A stratified random sampling technique was used to reflect the distribution of Corps members, half of whom were posted in the Ibadan Municipal area, while the reminder served the outlying Local Government Areas (LGAs).

Results revealed that health scientists, orthodox Christians, unmarried Corps members and those related to sickle

cell patients had higher mean knowledge scores for sickle cell disease. Regarding perception of the disease condition, most respondents (81.4%) said it was a serious problem. However, as regards perceived susceptibility of their own children, 68% of those untested for genotype and 53.5 percent of those already tested claimed their children would not be susceptible. Furthermore, among those untested, results showed that health scientists, male Corps members and orthodox Christians were most willing to undertake the test.

Based on these findings opportunities have been identified for improving the knowledge of this influential segment of Nigerian youth. Health Education at Universities and Polytecnics is therefore needed. In the meantime, health education in NYSC orientation camps can be strengthened.

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CERTIFICATION

I certify that this work was carried out by EREMUTHA Awasighe Francis of the Department of Preventive and Social Medicine, University of Ibadan, Ibadan.

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CHAPTER ONE

INTRODUCTION

The birth of children has been found to be the most sustaining factor in the institution of marriage (Arasomwan, 1987). Many couples view the birth and raising of children as the ultimate goal of family life, that is, the climax of successful marriage (Bamisaiye, 1974). In some families however, this happiness is shortlived by recurring indisposition or death of these much cherished offspring at infancy. Although the causes of infant deaths remain a mystery among most parents, some are usually attributed to cultural beliefs such as "ogbanje" and "abiku" - the children born to taunt their parents by dying young (Achebe 1965, Okri 1991). Recent advances in medicine however, showed that most infant deaths result from cell diseases (Whitten and Fischoff, 1974).

Sickle cell disease is the commonest type of gene influenced disorders. Others include phenylketonuria and myelomeningocele (Herrick, 1910; Serjeant, 1974; Powars, 1975) It occurs world wide and it is defined as the condition resulting from the inheritance of sickle haemoglobin (Hb SS) from both parents or sickle haemoglobin from one parent and another variant pathological haemoglobin from

the other parent (Ranney, 1954).

Although there are numerous types of sickle cell disease, three are common. Sickle cell anaemia is the most well known and is found in about one out of every 200 babies of West African origin (konotey-Ahulu, 1969) and about one in 300 babies of Afro-Caribbean origin (Serjeant,1971). The second type is the SC disease which is generally less common at birth , except in Ghana and is usually less serious (Konotey-Ahulu, 1973). The third type is sickle Beta-thalassemia which is the least common (Falusi, 1980; Jibril 1986).

Carriers of a single sickle cell gene are said to have sickle cell trait. They are as healthy as non-carriers, rarely having any health problems. For the three common types of sickle cell disease already mentioned, their carriers have haemoglobin genotypes AS,SC and S-Beta-thal. These are said to have resulted from the inheritance of the normal adult haemoglobin (HbA) from one parent and any of the variants from the other parent (Falusi, 1980; Anionwu 1986). Although people with the trait cannot later develop sickle cell disease, offspring of two such persons may inherit two sickle cell genes and develop the disease (Neel, 1949; Ranny, 1954; Weatherall, 1976).

The different kinds of sickle cell disease and the traits were originally found mainly in people whose families come from Africa, Asia, the West Indies, the Middle East and the eastern Mediterranean

(Herrick, 1910; Jelliffe, 1952; Esan 1970; Greenberg, 1970; Serjeant 1976). In recent times however, the disease had spread world wide through inter-racial marriages (Akinyanju 1989; Falusi 19 . It is now prevalent all over Africa, the Caribbean, Middle Eat, India, the Mediterranean and the United States of America (Serjeant, 1976; Oyejide 1982).

MAGNITUDE OF THE PROBLEM

The size of the sickle cell problem in Nigeria, like its population, is not static. Its alarming rate of growth is best appreciated when one pauses and considers the sheer number of expected birthe of affected children in one year. Dada (1988) and Akinyanju (1989) in separate reports observed that of the 5.76 million new born babies expected in Nigeria during the year 1990, 115,000 will have sickle cell anaemia, 22,000 will have SC disease while 1.25 million will have sickle cell trait. This estimation is based on haemoglobin S and C gene frequencies of 0.12 and 0.02 reported nationwide by Akinla (1972) and Fleming (1979) respectively.

In Nigeria, only the haemoglobin S and C are known to cause significant clinical problems. Of these, haemoglobin S is by far the commonest, and it is fairly evenly distributed throughout the country with reported heterozygote (AS) carrier rate of 25% in southern Nigeria, 19% to 32.6% in Northern Nigeria and over all

estimated average rate of 25% (Jellife and Humphreys, 1952; Walters, 1956; Fleming, Storey, Molineaux, Iroko, and Attal, 1979).

Among the Yoruba in Western Nigeria, haemoglobin C is common with a reported carrier (AC) rate between 5% and 7%(Lehmann, 1956; Nwokolo, 1959; Iroko, 1979). This is probably due to historical links and relative geographical proximity of the Yoruba people in Nigeria to the population of Modern Ghana with consequential inter-racial marriages (Esan, 1982). The incidence of haemoglobin C is 20% in Ghana, the highest so far reported in the world (Edington, 1956; Konotey-Ahulu, 1973). Adewuyi and Akintunde (1987) in a survey of 1,296 healthy children between age 1 month and 14 years in Ilorin, showed that 1.5% had haemoglobin SS, 1.3% had haemoglobin SC and 5.2% had haemoglobin AC. A similar survey of 1,022 pre-school children in Enugu showed that 11.6% had haemoglobin SS, none had haemoglobin AC (Kaine and Udeozo, 1981).

Earlier, Boyo Roberts and Lehmann (1960) had reported the existence of a small pocket of haemoglobin C in 5% of a small population of hillside Gwazo dwellers in Borno, Northern Nigeria. Akinkugbe (1978) in a study on the prevalence of anaemia among children age one to 10 years in Igbo-ora reported that 2.2% of 701 slides examined showed the presence of sickle cells, and this is similar to what has been reported by earlier workers (Hendrickse and Collard, 1960). Very recently in Benin City, Alumona (1989)

reported that sickle cell anaemia accounted for 25% of all children seen at the Benin Central Hospital Paediatric out patient during the year, 1988.

Apart from the rapid spread of the disease through inter-racial inter-tribal marriages, there is a high mortality rate associated with it. Jacobs (1957) and Abrahamson (1973) observed that sickle cell anaemia patients have less than 50% chance of attaining their twentieth birthday. The survival rate is even worse in developing countries like Nigeria where absence of efficient medical care disadvantageously affects the longevity of the patients (Anionwu and Jibril, 1986).

Fleming (1982) observed that the data from rural Nigeria on the longevity of sicklers suggest a situation where the unaltered environment (that is, minimal medical care for follow up of patients), is hardly compatible with the survival of SS patients beyond early childhood. In the report, it was noted that whereas SS patients were 30 per 1,000 in the new born population of a rural Nigerian district, the disease was absent in those more than 14 years in the same district. In fact, only one of the 8 patients expected to be in the one to 4 years age group was alive.

Earlier work by Mollineaux, Broger and Fleming (1976) in Africa revealed, also, that out of a total of 534 infants screened for major hemoglobinopathies in Garki, Nigeria, eleven babies were

found with SS, while 125 with AS were identified. On entry to the School programme at 5 years of age, when the same population was restudied, only one child out of 439 was found to have sickle cell anaemia but 133 were AS. The authors concluded that the babies with SS had died during early childhood and hence contributed disproportionate numbers to the high infant mortality in Nigeria. No doubt, the situation was probably worse than reported in the above epidemiologic investigations. For, as the report also noted, documentation of the incidence of sickle cell anaemia and its effect on childhood mortality actually started after Fleming and other investigators have commenced the cord blood hemoglobinopathy surveillance programme.

The socio-economic implications of this high death toll are grave. With low life expectancy, relatives and parents of sicklers suffer severe emotional stress because of unfulfilled hopes. The birth of a child, which is supposed to be the family's crowning glory, becomes for them mere physical agony devoid of promise (Achebe, 1964). Most marriages resulting in sickle cell children suffer unhappy parenthood, instability and sometimes divorce after all efforts to produce a healthy living child fail (Bamisaiye et al, 1974; Okunade 1982).

On the society, the impact of the disease is no less severe. Leigh (1987) observed that sicklers are a group of invisible

handicaps whose survival depend on the amount of sympathetic exemptions that society is willing to make on their contribution to community growth and development. Sufferers occupy the sick role most of the time (Milner and Desforges, 1987) and thus need extra attention to survive. This poses both economic and social strain on families and communities. Even those few who survive may be permanently disabled and unfit for services to society (Abrahamson, 1973) and thus unable to pay society back for the assistance received.

PURPOSE OF STUDY

Despite the high sickling rate in Nigeria, Akinyanju (1989) reported that awareness of the Nigerian populace about sickle cell disorder is, by anecdotal accounts, very low. In rural areas especially, high mortality among infant patients is closely associated with the "abiku" and "Ogbanje" phenomenon. As a result, Akinla (1972) and Fleming (1982) observed in separate reports that 70% of sickle cell babies in Nigeria die undiagnosed before attaining the age of five years. This is because of the urban concentration of medical facilities needed for diagnosis and treatment of the disease.

To date, national efforts to increase awareness about sickle cell disease have been sparse. The recent requirement for haemoglobin genotype testing as a pre-condition for issuance of a

driver's license is a potentially good step. On the local level, there are sickle cell clubs (Lagos, Ibadan, Ilorin , Benin) who have conducted awareness campaigns in schools. Generally, a concerted and coordinated educational effort is lacking.

In the main, health scientists and researchers are unanimous on the opinion that genetic dilution through appropriate choice of spouse is and will continue to remain the most viable option in the quest for solution to the problem of sickle cell disease (Falusi, 1989). The selection of the appropriate spouse in a society like Nigeria with high ignorance and high illiteracy rate depends to a large extent on the quality of enlightenment campaigns undertaken for the purpose (Borrofice, 1981; Mathur, 1983; Akinyanju, 1989). Such campaigns become more effective when educated and influential members of the society are used as change agents and when facilities for testing are available.

The National Youth Service Corps (NYSC) members are an influential group that enjoys extensive community cooperation, respect and peer identity. By virtue of their educational attainment, age and location, they may serve as change agents to society. As vanguards of modern society, they have the potential to influence others by what they know, say or do. Their engagement during the service year as teachers, counsellors, extension workers and health personnel in all nooks and corners of the country

enhances their contribution to society as change agents.

It is this unique and privileged position of the Youth Corps members that necessitated the need to assess their knowledge and perception about sickle cell disease and to document, through this study, their willingness to undertake the appropriate preventive measures.

THE NATIONAL YOUTH SERVICE CORPS SCHEME

Nigeria is a country whose colonial history and experience in the immediate post-independence era are characterized by ethnic loyalties, mutual group suspicion and distrust (NYSC, 1988). As a developing country, Nigeria is further plagued by the problems attendant to underdevelopment, poverty, mass illiteracy, shortage and uneven distribution of high skilled manpower and a woefully inadequate socio-economic infrastructure. Aware of the intractable nature of those problems, government and people set for the country fresh goals and objectives aimed at establishing a strong, united and self-reliant nation. It was in realization of these goals and in recognition of the role of youth in the sustenance of society that the National Youth Service Corps Scheme was established by Decree 24 of 22nd May 1973 (NYSC, 1983).

The objectives of the scheme are as follows:

a. To inculcate discipline in Nigerian Youth by instilling in them a tradition of industry at work and of patriotic and

loyal service to the nation,

b. To raise the moral tone of our youth by giving them the opportunity to learn about higher ideals of national achievement and socio-cultural improvement,

c. To develop in our Youth attitudes of mind, acquired through shared experiences and suitable instruction, which will make them more amenable to mobilization in the national interest,

d. To develop common ties among our youth and promote national unity by ensuring, as far as possible, that youth are assigned to jobs in states other than their state of origin and away from their geographical, ethnic and cultural background; that each group assigned to work in each state, is as representative of the country as possible; that the youth are exposed to the modes of living of the people in different parts of the country with a view to removing prejudices, eliminating ignorance and confirming at first hand, the many similarities among Nigerians of all ethnic groups,

e. To encourage members of the Corps to seek, at the end of their Corps service, career employment all over the country, thus promoting free movement of labour,

f. To induce employers, partly through their experience with Corps members, to employ more readily qualified Nigerians irrespective of their states of origin, and

g. To enable our youth to acquire the spirit of self reliance (NYSC Decree, 1973).

The decree made it mandatory for all Nigerian graduates from Universities and Polytechnics both at home and abroad to participate in the one-year National Youth Service Corps Scheme. However, section 1 of the NYSC Act, 1985, exempted the following categories of diploma or degree holding graduates: those over 30 years; police and armed forces personnel; prisoners; persons conferred with national honours; persons certified insane by a Government psychiatrist and persons certified incapacitated but excluding the blind, deaf, crippled and dumb whose incapacitation does not deter them from academic pursuits.

The Youth Service is divided into two major parts. The first four weeks consists of an orientation exercise in camps during which members undergo drills, physical exercises, lectures and leadership training activities. The second and greater part of the service year involves deployment of members to work in areas relevant to their fields of study or in the National interest. In addition, a one day per week community development programme takes place in which corps members undertake projects that are the felt need and of developmental importance to the communities which they serve. The second part of the service year follows immediately after the orientation exercise, and corps members during this period are

posted to primary assignments specified in the Decree establishing the scheme. Such Primary assignment postings take corp members to all government departments and statutory corporations suitable for new graduates, the private sector of the Nigerian economy, hospitals, road construction, farming, teaching, food storage and eradication of pests, rehabilitation of destitutes and the disabled, development projects of local councils, social and economic services, and other undertakings and projects as the Head of the Federal Government may by order determine.

A participatory management scheme operates. Corps members are involved in administration of the Scheme at the State and Local Government levels through their role as Corps liaison officers. Corps liaison officers are responsible for linkage between the Corps members and the State NYSC secretariats. The arrangement complements the efforts of NYSC Zonal supervisors (NYSC, 1983).

The achievements of the scheme especially in the attainment of its primary objectives are visible. In spite of societal pressure for concessional posting, rejection of rural postings by some Corps members, evasion of service, abscondment from service, poor camping facilities, and a difficult employment prospect for graduates, the NYSC is no doubt, in the vanguard as an instrument for labour mobility. Even socio-economic development and the promotion of inter-ethnic understanding for national unity are some of the many

benefits of the scheme (NYSC, 1988).

In the area of manpower supply and labour mobility, the scheme has made major achievements especially in the realms of health and education. A study by the Research Division of the NYSC (1986) revealed that all eleven old states that provided data enjoyed the services of no less than 38 corps health personnel. The report also revealed that there was hardly any secondary school in the country where at least two or three corps members were not part of the teaching staff. In 1988, another survey by the same division also showed that of the 350,000 corps members so far mobilized during that year, over 65% were assigned to secondary schools and other educational institutions (NYSC, 1986).

In his congratulatory message to the NYSC on the occasion of the celebration of its 15th Anniversary, His Highness, Oba Lamidi Olayiwola Adeyemi III, the Alafin of Oyo commented

> "Let us take the case of teachers of specialized and technical subjects like science, mathematics and English Language who were mainly concentrated in urban centres. The result was that schools in the villages and remotest rural areas never had the opportunity of access to facilities of teaching sciences and mathematics because no teacher of these subjects would love to leave the cities for the villages. But because the NYSC postings

are more or less like military postings one is happy to see today science and mathematics teachers in the remotest areas of our villages. That could not have been so but for the NYSC Programme" (NYSC, 1988).

Apart from their primary assignments many corps members, irrespective of their areas of specialization, organize extramural, adult education evening classes as well as literacy campaigns and science clubs, for example in Nyanya, Kuru and Kabochi in Abuja (NYSC, 1984). In Gongola State, an education committee was constituted by 1986/87 service year Corps members with the aim of raising the educational standards of students in the state by organizing extramural classes, career guidance and counselling programmes, intra-school debates, essay writing and quiz competitions (NYSC, 1988).

Regarding the contribution of the scheme in the area of health care provision, evidence abounds in many states where Corps members have initiated community based health centres for the benefit of their host communities. A few examples of Corps member initiated projects in the above regard includes the establishment and running of dispensaries at Agyanan Village in Abaji Local Government and at Rimaye in Dutein-Ma Local Government Area of Kaleina State (NYSC, 1986). Others include free health consultative services organized by Corps health personnel in Lagos, Bauchi, Kwara and Borno States

(NYSC, 1989).

During the 1984/85 service year, the health team that arrested the spread of Guinea Worm in Kankan, Asa Local Government Area of Kwara State, was led by a Corps member (NYSC, 1988). In Borno State, the eleven comprehensive health centres located in the rural areas are manned by Corps doctors. No wonder, an opinion leader in Konduga, Borno State, once said "The greatest thing that has ever happened to our village is the presence of NYSC doctor with us" (NYSC, 1989).

The former Niger State Governor Colonel Lawan Gwadabe (NYSC, 1990) on the occasion of the opening of the NYSC 33rd bi-annual conference held in Minna, in June 1988, in recognition of the past performance of the Corps doctors gave the following promise:

> "Any Corps doctor deployed to serve in Niger state has been assured of free comfortable accommodation, free transport, free medical care for himself and family during his service year. Above all, he is equally to be offered automatic post service and pensionable employment in addition to other fringe benefits".

With respect to the scheme's promotion of inter-ethnic understanding, integration and national unity, much has been achieved. Even though it is difficult to quantify the rate of

attitude change, there have been testimonies by Corps members, their employers, as well as the operators of the scheme on how numerous Corps members have learnt to understand and integrate better with their host communities during their service year (NYSC, 1987).

In the area of agriculture, NYSC has successfully established and cultivated arable farms in all former 21 states of the Federation and the Federal Capital Territory. Along with crops, successful poultry farms have been established in Lagos, Ondo, Oyo and Niger States as wells as pilot fish farms and rabbitory in Ondo, Rivers, former Bendel and Oyo States (NYSC, 1983).

For infrastructural and technological development, almost all communities in the Federation have, in one way or the other benefitted from the roads, market stalls, bridges, art works, geological and hydrogeological maps and a host of other projects too numerous to mention (NYSC, 1988). All these, are in addition to such specialized services rendered by Corps members including the Corps Legal Aid Scheme in which Corps members render free legal services to the public. Corps members also left their marks in their service locations through group and individual activities aimed at beautifying and improving communities (NYSC, 1989).

It is no wonder that the influence of NYSC members in the communities where they serve is massive. In the recent years, Corps members have been known to participate actively in getting people at

the grassroots level well informed on social, health, economic and development programmes (NYSC, 1986).

BACKGROUND OF THE STUDY

Being based in the capital of Oyo State during the study, the researcher decided to focus on NYSC members located in that state. At the time of the project (February, 1991) Oyo State was the second largest of the then 21 states of Nigeria, and its capital, Ibadan, was the largest indigenous African town in south of the Sahara. The state then was made up of 42 Local Government Areas (LGAs) amongst which Ibadan Municipal Government (IMG) was the largest and most developed.

The area that constituted the state at the time of the project was bounded by Ogun State to the south, Republic of Benin to the west, Kwara State to the north and Ondo State to the east. It covered an area of 27,600 square kilometers and had a population of 10,044,826 (1990 estimate). The principal towns in the state were Ibadan (the state capital), Ogbomoso, Osogbo, Ile-Ife, Ilesha, Saki, Iwo, Iseyin, Ikire, Ede and Gbongan (Ministry of Local Government and Information Ibadan, 1990).

The state lay between the high forest zone in the south, which is mainly tropical rain forest, and the savannah in the north, which is a belt of grassland. The indigenes of the state are the Yoruba who are mainly christians and muslims although some still belong to

Indigenous religions. The state is mainly agricultural and a major cocoa and palm oil producer. In addition, food crops such as yam, rice, maize, beans, cassava, plantain, banana and fruits are agricultural staples.

There is an urban tendency among the people of the state because of the commercial nature of the principal towns in the state. This also accounts, for the large number of the non-indigenes from neighbouring states and countries found in the state. Notable among the immigrants are the Ghanians and the Asians. The indigenes of the state have a common culture and their various festivals are similar in organization and celebration (Ministry of Local Government and Information, Ibadan, 1990).

The result of twenty-one years (1955-1985: excluding the military regime) of free primary education is the establishment of numerous primary, secondary and tertiary schools within the state. The total student enrolment in the state during 1990 was 953,421 for primary schools and 390,587 for secondary schools. The state had five tertiary schools, nearly three hundred colleges and a little more than four hundred primary schools (Oyo State Ministry of Education, 1990).

Apart from the villages and hamlets, all cities and towns in the state had adequate supply of electricity and pipo borne water. However, the condition of access roads was especially poor during

the rainy season. Ibadan Municipal Government area (IMG) is the most developed LGA in the state and had the largest concentration of educational, health and industrial establishments. It is no surprise therefore, that half or more of total Corps members deployed to the state each year were posted within the IMG, while the remainder served in the other 41 LGAs. More details on the distribution of Corps members in the state are found in Chapter Three.

In the next chapter, the author reviews literature relevant to sickle cell disease and preventive health behaviour. Chapter Three describes the methodology used in collection of data for this study. The instruments used are described including their validity, reliability and mode of administration. The major findings of the study are presented in Chapter Four, while Chapter Five consists of discussion, conclusions and recommendations. Of interest to readers are the appendices that contain definitions of terms used in the study and samples of the study instruments.

CHAPTER TWO

LITERATURE REVIEW

This chapter covers the nature and extent of sickle cell diseases and the problems faced by families having children with the condition. The section also deals with issues of screening and counselling as related to the disease. Finally, the chapter is concluded with explanations of preventive health behaviour in relation to the concept of the Health Belief Model - the conceptual framework utilized in the study.

NATURE AND EXTENT OF SICKLE CELL DISEASE

A controversy still exists as to who the first person to discover the sickle cell disease. Flint (1904) however subscribed to the popular belief that Dresbachs' (1904) report of "a peculiar anomaly in human red blood corpuscles" of a mulatto who died later was the first case of sickle cell disease available in the literature. Dresbach (1905) however maintained that the patient in question did not die as a result of anaemia and hence refuted what would have been a consecration of medical history in his credit. Nonetheless, Sturgis (1948) stated that since the subject was a mulatto and had died of cardiac disease at a young age, he might have suffered from sickle cell anaemia.

In the face of these controversies, the "peculiar elongated and sickle-shaped red blood corpuscles in a case of severe anaemia" reported by Herrick (1910) appeared to be the first clear case of sickle cell anaemia in literature (Huntsman and Lehmann, 1966). Herrick (1910) enumerated the phenotypic and clinical presentations observed in the patient. The same of which were reported about another case by Washburn (1911).

Cook and Meyer (1915) also reported another case which by now they believed was a familiar clinical entity. But perhaps, the boldest and most specific impression was that expressed by Mason (1922). On the basis of an extensive study on the human blood under low oxygen tension by Emmel (1917), Mason reported the fourth case and called it sickle cell anaemia without reservation.

As soon as sickle cell anaemia became accepted as a clinical entity, reported cases began to appear in the literature. Within the last 44 years precisely, many contributions have been made in every field concerned with sickle cell anaemia and its varied manifestations. Most outstanding among these contributions are those on heredity by Neel (1949; 1951), haemoglobin variant by Panlin (1949; 1950) and the molecular basis of the error in sickle cell disease by Ingram (1957; 1958) and Ingram and Stretton (1959).

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Today, sickle cell disease has become recognized as an international health problem and many international institutions and associations have become established with the hope to finding lasting solutions to this hereditary haematologic disorder.

Etiology

Paulin (1949) was the first to associate sickle cell disease with an abnormal haemoglobin. According to him, since low oxygen tension leads to sickled erythrocytes and local anoxia causes damage to organs, a molecular abnormality of haemoglobin must be responsible. This was confirmed later by the difference observed in the electrophoretic mobility between haemoglobin obtained from sickle cell anaemia patients and that obtained from normal blood cells. He therefore concluded that sickle cell anaemia haemoglobin (Hbs) was different from adult haemoglobin (HbA).

Schroeder and Matsuda (1958) observed that normal adult haemoglobin (HbA) consists of four polypeptide chains, two alpha (α) chains and two beta (β) chains. The amino acid sequence was worked out by Braunitzer (1964) during which he found the alpha chain to contain 141 amino acid residues, while the beta chain contains 146 amino acid residues. He reported that each chain is reversibly bonded to heme, a compound of one iron atom combined with a molecule of porphyrin.

Apart from the primary structure of the polypeptide chains

(the sequence of amino acids) each chain has a secondary structure by coiling in to helical structures in several places (Giblet, 1969). It was additionally noted that the four chains in a haemoglobin molecule are further folded in characteristic ways that confer on them a three dimensional structure referred to as the tertiary structure of haemoglobin (Cuills, 1962; Mulrhead and Perutz, 1963) thus, changes affecting the primary, secondary and tertiary structures and consequently the physical properties may bring about measurable changes in the red cells, which vary from moderate to severe (Gilbert, 1969).

In normal adult red cells, two main types of haemoglobin, A and A2, exist. In most individuals, the proportion is 97% A and 3% A2 (Kunkel, 1957). Haemoglobin A2 consists of two alpha and two delta chains (α_2 and σ_2) which replace two beta (β) chains in haemoglobin A (Ingram and Stretton, 1961). The alpha chain in the two haemoglobin types are identical, while the delta chains differ from the beta chains in terms of ten differences in their sequence (Hill and Kraus, 1963; Jones, 1964).

Fetal haemoglobin, which is the major haemoglobin component during fetal life, has a concentration of less than 0.5% after the first few days of life in normal individuals (Giblet, 1969). Fetal haemoglobin (HbF) consists of two alpha chains identical to those of Hb A and Hb A2, and two gamma chains which differ from

beta (β) chains in 39 amino acid residues (Schroeder, 1963).

After Paulin (1949) had shown that haemoglobin S is different from haemoglobin A, Ingram (1958) observed that peptide maps of the beta polypeptide chain of Hb A differ from those of the maps of the beta chain of HbS by one peptide. Ingram (1959) stated that the single peptide difference is due to the substitution of one amino acid of a different charge for the normal amino acid, thus accounting for the different mobility of the polypeptide chains. According to him, HbS results from the substitution of valine for glutamic acid in position six of the beta chain of HbA.

A host of other abnormal haemoglobins were later discovered. In some, the beta chain is normal while in others, it is the alpha chain. Table 1 presents a list of some abnormal human haemoglobins and the corresponding amino acid substitution of each. Harris (1950), Mitchison and Perutz (1950), reported that under low oxygen tension, HbS forms spindle-shaped crystals, a phenomenon which has been associated with the intramolecular changes brought about by the substitution of the valine for the glutamic acid.

The HbS molecule, as a result of these intramolecular changes, undergo a "head to tail" stacking with formation of filaments. These filaments may twist together into hollow

TABLE I

SOME HUMAN HAEMOGLOBIN VARIANTS AND
THE CORRESPONDING AMINO ACID SUBSTITUTION

Haemoglobin	Abnormal Chain	No. of Sequence	Change
S	β	6	GLU-VAL
C	β	6	GLU-LYS
D-Punjab	β	121	GLU-GLY
E	β	26	GLU-LYS
D-Ibadan	β	87	THREO-LYS
G-Accra	β	79	ASP-ASN

[SOURCE: Ingram, 1959]

microtubules forming the basis of a crystalline structure. Murayama (1965; 1966) and Murayama and Stephens (1965) noted that sickling does not occur when haemoglobin S is fully oxygenated because beta chains move towards each other thus increasing the distance between beta and alpha chains of adjacent molecules. This is due to the fact that stacking, which causes sickling, does not occur when beta and alpha chains of adjacent molecules are far from each other.

Mode of Inheritance

Cook and Meyer (1915) were the first to recognize the familiar nature of sickle cell disease. Eight years after their report, Huck (1923) suggested that the disease might be inherited as a mendelian dominant trait. They had inferred that the gene had a variable expression which was more pronounced in some individuals producing sickle cell anaemia and weaker in others, resulting in what is now referred to as sickle cell trait.

The victims of sickle cell disease are born with it just as they are born with other characteristics such as eye colour, hair texture and height. It is inherited from both parents through substances called genes. The gene for haemoglobin inherited from both parents determines whether the child will suffer sickle cell disease or not. Everybody inherits two genes, one from each parent. There are four most important genes involved in sickle

cell disease and trait. The most common gene is haemoglobin A (HbA) which is the normal haemoglobin. Others are sickle haemoglobin S (HbS), haemoglobin C (Hbc) and beta-thalassemia (Beta-thal) which are abnormal haemoglobins. If a child inherits two genes for haemoglobin A (HbA), the child will have normal haemoglobin (HbA). However, if the child inherited one gene for haemoglobin A (HbA) and one for sickle haemoglobin (HbS), the child will have sickle cell trait (HbS). Individuals with sickle cell anaemia inherited one sickle cell gene (HbS) from each of their parents resulting in (HbSS), the most common kind of sickle cell disease.

Neel (1949) put forward a different hypothesis for the etiology of the disease. He proposed that sickle cell anaemia was determined by homozygosity for a recessive gene inherited from both parents. This was confirmed by Beet (1949) after studying a Bantu family in which a child died of sickle cell anaemia. Six living siblings in this family had the trait as did the two parents.

Through subsequent work by Paulin (1949) and Neel (1951), it was established that sickle cell anaemia is determined by homozygosity for a recessive mendelian gene and that the sickle cell trait is an heterozygote condition, the transmission of which is summarized in Table 2.

Table 2

Transmission of Sickle Cell Anaemia in Different Mating Types

Mating Type (Hb Genotype)	Percent of Offspring		
	AA	AS	SS
AA x AA	100	-	-
AA x AS	50	50	-
AA x SS	-	100	-
AS x AS	25	50	25
AS x SS	-	50	50
SS x SS	-	-	100

A = Dominant Gene for Normal Haemoglobin A.

S = The Recessive Allele Producing Haemoglobin S.

[Source: Neel, 1951]

Distribution and Frequency of the Gene

Since Herrick (1910) described sickle cell disease in a Jamaican, evidence of its presence in numerous blacks has been confirmed. The epidemiological survey of the disease by Livingstone (1979) showed that although it occurs predominantly in black populations, it is more widely distributed among the world's population than those with African ancestry. He noted that its distribution in the old world is almost in accordance with the distribution of falciparum malaria with the exception of South East Asia.

The World Health Organization (1966) reported that in Africa, the sickle cell gene (Hbs) is distributed in a broad equatorial belt extending from ocean to ocean. It's distribution is said to be limited to the north by the desert and the Ethiopian highlands. Southward, it extends approximately to the river Kunene in the west and to the river Zambezi in the East.

In India, the gene has been reported with highest frequencies among the Konds of Bastar and especially among the hill tribes of the Negroid Veddoids of Southern India. In the Middle East, populations with sickle cell gene are scattered especially in Iraq, the Delta of the Tigris and Euphrates Rivers, and into the Persian Gulf. Isolated areas in this region with severe endemic malaria also have high frequencies of the disease.

but in Arabia, sickle cell is not found in the pure Bedouins. Similar cases of sickle cell gene in "whites" of the Mediterranean have been noted (Odgen, 1943; Sights and Simons, 1931; Hadan and Evans; 1937).

Sickle cell trait and disease are as prevalent in the United States today as in the past. One in 12 blacks in America carries a gene for sickle haemoglobin, and one in every 400 - 500 black newborns has sickle cell anaemia. However, they are not the only ones who can be afflicted. Genes for sickling and other abnormal haemoglobins are sometimes carried by persons of Greek, Puerto Rican, Spanish, Portuguese (especially from the Cape Verde Islands) and Italian ancestry (Wethers, 1978).

Thus, sickle cell disease which for many years appeared to be virtually confined to the black population of equatorial Africa and the descendants of West Africans in the New World has gradually become apparent among other races. It is recognized that the gene is widespread and not racially linked, the factor common to its distribution being falciparum malaria, because under malarious conditions, the heterozygote for HbS enjoys better health and a consequently greater chance of transmitting his genes (Serjeant 1988).

PROBLEMS FACED BY FAMILIES OF SICKLERS

Marriage, according to Bell (1972; 1984), is the basic unit

upon which the family is generally built. This unit entails a rearrangement of social structure through the institutionalization of persons and relationships (Radchiffe-Brown, 1950). Through marriage, new social relations are created, not only between the husband and the wife, but also, in a great many societies, between the relatives of the husband and those of the wife, who on the two sides, are interested in the marriage and the children that are expected to result from it (Arasomwan, 1987).

One major factor that determines marriage happiness and stability is the survival of offspring resulting there from (Marrinin, 1976; Mueller and Hallowell, 1977; 1986). This is because the typical African society is pronatal (Morris, 1961) and usually adduces superstitious explanations for both morbidity and mortality in children (Achebe, 1968).

Very little is known about the science of inheritance by the general populace, although some people acquire some knowledge of it when they are faced with the socio-economic and emotional problems associated with the presence of a sickler in the family (Leai, 1985).

The socio-economic and emotional problems caused by the presence of sicklers in families are grave particularly because of the high death toll observed among the patients (Abramson, 1973; Diggs, 1973; Konotey-Ahulu, 1973; Fleming, 1979; Anionwu, 1986,

Akinyanju, 1989). Most marriages resulting in the birth of sicklers undergo gradual disintegration because of excessive demands on the family's finances following the recurrent hospitalization of the sickler (Bamisaiye, 1974; Okunade 1982). One common feature of such marriages is unhappy parenthood and sometimes divorce. This is because survival of the children is the second major factor which sustains the marriage institution, followed closely by sexual consummation (Mair, 1969).

Another social problem which often affects families with a sickle cell patient are the feelings of shock, guilt, annoyance, fright and distress, which follow the disclosure of the fact that their child has inherited the sickle cell disease. Such feelings, according to Anionwu and Jibril (1986) and Anionwu (1989) are both common, especially if neither of the parents is aware of a history of illness in their family. In other cases, parents regard the child's illness as punishment for something they have done. Brothers and sisters may also feel guilty because they are healthy and "cannot take share" of the pain (Phillips and Joseph, 1976).

It is quite common for one partner to blame the other, or to think that it must be his or her side of the family that is passing on the illness without the knowing that sickle cell disease must be inherited from both parents. Fleming (1982) also observed that parents' inability to predict when a child with

opinions These interventions are often dangerous and expensive. Herbs and roots, not fully tested, may shorten the lives of certain individuals especially sicklers (Scott, 1976). Lesi (1985) reported that there is a tendency for parents of sicklers to "shop" around for a cure. Often, they are deceived by unscrupulous people who claim they have the cure through the use of herbs, roots, special soaps and supernatural means such as invocation of the bad spirit out of the child's body.

The psychological effects and emotional disequilibrium which parents suffer are equally severe. The unending patronage at both traditional and orthodox healing homes results in serious emotional distress. The parents live in perpetual fear as they run from one spiritual home to another for assurance that the patient would get well and in fact survive the death threat (Leigh, 1987). Furthermore, they suffer from fear and anxiety about the fate of subsequent pregnancies (Olatawura, 1976; Konen and Schmidt, 1978).

The psychological strain placed on families by the birth of sicklers is especially pathetic when one realizes the high premium placed on the birth and survival of children in the typical African Community. The situation is particularly bad if the sickle cell child is delivered by one of the many wives in search of a child in a polygamous family (Bamisaiye, 1974; Leigh

1987).

Despite the serious negative effects of the sickle cell phenomenon on the stability and happiness of marriages and families, a majority of youth and parents alike remain poorly informed or even completely unaware about the disease. At the moment, deaths of infant and child sickle cell patients continue to be attributed to cultural beliefs such as "ogbanje" and "abiku," the children born to taunt their parents by dying young (Achebe, 1964; Ebomoyi, 1988). This is perhaps the basis upon which Falusi (1989) advocated for concerted efforts towards public enlightenment about the disease and pre-marital counselling among the youth. This can be achieved only through well coordinated and organized screening and counselling services (Oberteuffer, Harelson and Pollock, 1972; Shortell and Richardson, 1978; Falusi, 1988; Ebomoyi, 1988).

SCREENING AND COUNSELLING SERVICES

It is easy to assume that people who appear to be fit are also healthy. This is not always true. Diseases such as tuberculosis, lung cancer and sickle cell trait can be present in an individual and yet that person appears healthy (Stride and Stride, 1986).

To achieve the aim of genetic counselling, the diagnostic procedure must be as accurate and possible. In screening for

genetic disease, minimal procedures include Alkaline haemoglobin electrophoresis with the use of the sickle or solubility test to confirm the identity of a HbS band (Barnes 1972; Giorgio and Boggs, 1974).

The most appropriate age at screening is controversial, the ages at which the population are most accessible not always being those at which this knowledge is most appropriate for genetic counselling. Most of the population is readily accessible at birth, yet the information on the sickle cell trait among new borns is unlikely to be used for 20 years, although of course, detection of a newborn with the sickle cell trait serves to identify one parent, and possibly two at risk of having another child with sickle cell disease. The population is also again accessible at school and before marriage (Serjeant, 1985).

The target population for screening has also been controversial, and the educational programme must be directed at all sectors of the population carrying the sickle cell gene. Also, education about the significance of the results obtained by screening is as essential as the screening exercise itself (Serjeant, 1985; Aninowu, 1986; Akinyanju, 1989; Falusi, 1989).

Screening may be for prevention of sickle cell diseases, as is case for trait identification, particularly in adults. The result of such screening guides the content and direction of

sickle cell prevention counselling. Genetic counselling, rather that counselling in health problems, is the major reason of screening black people for sickle cell trait. In the process however, a few cases of sickle cell anaemia may be detected. Advocates of the preventive sickle cell policy feel that these people have a right to know about their condition, so that they can make up their own minds about marriage and raising a family.

Preventive screening exercises and their results enable the counsellor to set aside time to explain to patients (Amonwu and Jibril, 1985). It also provides an opportunity for reassuring patients that the trait in no way impairs their health. This avenue can also be used to explain to carriers that they could experience sickling in the renal capillary bed and that this requires thorough investigation. But most importantly, the patient should be told that he could have sicklers or carriers as children, depending on the Hb genotype of his spouse (Falusi, 1989).

The other type of screening which has gained medical publicity is screening for early detection of infant patients. Cord blood screening for sickle cell anaemia is gaining favour. New York State (U.S.A.) has a mandatory screening law. New born screening according to Serjeant (1985) lowers the mortality rate since the first disease related episodes can be fatal. Thus, this

type of screening is geared towards identifying patients to manage and is not immediately geared to prevention, unless it guides the parents in future childbirth plans (Pearson, 1989; Powars, 1989; Vichinsky, 1989).

Apart from the law in New York State, many medical centres and other states in the U.S.A. have similar statutes for screening both black and white neonates. However, due to their seemingly coercive nature, these laws have been repealed. In many instances, the need for screening is obviated by the fact that both parents have previously been tested and found not to have the trait. If only one parent has the trait, there is no need to screen the infant for disease, although he may have trait (Grover, 1989).

Screening is encouraged among all who do not know their Hb genotype and for children of couples where one parent has the sickle gene and the other has either the sickle gene or that of C or beta-thal (Walker 1989; Audrey, Scott and Agatisa, 1989).

In Nigeria, very few people have access to a sickle cell screening facility. This is because the 17 sickle cell centres equipped for the exercise are concentrated in the urban centres. The result is that the 17 centres are usually crowded and yet under-staffed. Generally the centres are located in teaching hospitals. Private hospitals and laboratories also conduct the Hb

genotype screening at a rate of 35.00 to 50.00 Naira, while the test costs fifteen Naira in government hospitals, making patronage at such private hospitals low (Lesi, 1985).

Three main tests are available in each of the testing centres. These are the sickle test, the solubility test and haemoglobin electrophoresis. The sickle or solubility test shows whether an individual has sickle haemoglobin in his blood. It is carried out by mixing one drop of blood with a reducing agent on a glass slide covered by a smaller piece of glass and sealed so that oxygen does not reach the red blood cells in a drop. This test does not distinguish between sickle cell disease and sickle cell trait. Also, it cannot be used to test babies under one year, as the results are not reliable (Anionwu and Jibril, 1986).

The solubility test depends on the principle that haemoglobin S is less soluble than haemoglobin A when mixed with certain chemicals in small glass tubes. It can serve as a guide in distinguishing between sickle cell anaemia, sickle cell trait and the normal haemoglobin. It has similar drawback to the sickling test, and haemoglobin electrophoresis must be carried out to confirm the result (Giorgio and Boggs, 1974).

Haemoglobin electrophoresis is the most reliable test for the sickle cell disease. It indicates exactly the type of haemoglobin that the individual has when the chemical used in

changed from one type to another (Barnes, 1972).

The absence of a cure for sickle cell disease, at the moment, makes counselling of screened individuals mandatory. Fraser (1974) listed the following as the major goals of genetic counselling:

a. establishing the risk of recurrence of the disease.

b. interpreting the risk in meaningful terms.

c. aiding the counsellee to weigh the risk and formulate a plan of action

d. follow up the counselling to reinforce the risk figures and estimate its effects on the counsellee.

Essentially, the process entails giving advice or information on inherited disease with particular reference to why and how it occurs. The way the disease is identified and the means to prevent it are also of great importance in counselling. Effective genetic counselling should involve two or all of the following.

a. the parent from whom the disease is derived,

b. the patient or potential patient if any, and

c. the counsellor.

In a condition such as sickle cell disease, counselling must be held in plain language, unrushed, and should aim at educating the people about various aspects of genetic disease. It is also

important that the counselling environment be one of friendliness, familiarity and sympathy, since these are essential parts of the management of the condition (Leal, 1985).

In the United States of America, a great deal of experience has been gathered over many years in the area of genetic counselling. The U.S.A. is one country that has the advantage of adequate government funding and also the convenience of addressing the comparatively literate black population in only one language, that is English. In the U.S.A., one in ten people of African origin carries the trait of sickle cell, and one in 400 blacks suffer from sickle cell anaemia (Schmidt, 1974; Heafner, 1974; Shortell and Richardson, 1978).

The objective of counselling in the United States and indeed all well meaning countries, is to eradicate the disease in the future using the prospective counselling approach. In this approach, which is perhaps the most efficient and effective, counselling involves the testing and talking to every black, old and young (Leal 1985; Ebomoyi, 1988). It however, involves huge financial outlays and carries the potential of political misinterpretation, especially when marriage choice is statutorily limited between certain individuals. Also, in communities where the people are sensitive and intensely aware of the colour of their skin, as in the United States, selective marriage may brin

about conflict between races (Kenen and Schmidt, 1978; Olatawura, 1979).

Most African countries, including Nigeria, lack the "political will" to undertake prospective counselling as a sickle cell preventive measure, even though they harbour the largest percentage of the HbS gene world wide (Lesi, 1985). Only seventeen government hospitals in Nigeria have the means to do a simple haemoglobin electrophoresis to determine a person's genotype. The majority of these are urban located and in fact are mainly teaching and specialist hospitals. In these centres, prospective counselling, which addresses only families identified to be at risk of producing sickle cell disease patients, is adopted by the nurses and doctors in charge of the sickle cell clinics (Anionwu, 1985; Anionwu and Beattie, 1986; Falusi, 1988; Ebomoyi, 1988; Akinyanju, 1989). Although, prospective counselling has the advantage of addressing the peculiar problems of each at-risk family identified at the sickle cell clinic, and has enjoyed relative success over the years at the Lagos University Teaching Hospital, it nevertheless, lacks the capacity to reach a larger audience as does retrospective counselling. Prospective counselling employs specialized laboratories, staff, and sophisticated communication gadgets to reach churches, supermarkets, schools and social centres (Anionwu, 1985; Ebomoyi,

1988).

By and large, screening for the trait as prevention against sickle cell disease may cause harm if it is not possible to follow up with counselling, as those with the disease or trait may suffer stigmatization, job discrimination and be rated as high risk by insurance companies (Olatawura, 1976; Kenon and Schmidt, 1978; Akinyanju, 1989; Whitten, 1989).

In order to avoid the untoward effect of a screening programme that lacks the counselling component, Luzzatto (1975) and Anionwu (1985) recommended that, in addition to making counselling compulsory in screening centres, both retrospective and prospective counselling approaches should be adopted at every centre . This is particularly important because of the estimated risk of affected offspring which shows that a couple carrying the AS trait each has a 25% chance of having an SS child. On the other hand, if they already have one or more children with sickle cell disease, the risk of reoccurrence is still 25% because chance has no memory (Lehmann, 1985).

Counselling therefore, must assure that carriers of the sickle cell trait are made aware of this fact before making a decision on marriage partners. Married couples too, if they know, would be able to decide on the number of children they want, as well as be able to prepare for the troubles of parenting

sicklers. This is why attempts to control the disease should aim at letting the public know what the disease is. However, this must be accompanied by proper education on the repercussions of being saddled with a sickling child even before marriage is embarked upon by carriers, and to know what to do about screening for the disease and where to go for counselling.

PREVENTIVE HEALTH BEHAVIOUR

Organized health education activity is based on the desire to intervene in the process of development and change in such a way as to maintain positive health behaviour or to interrupt a behavioural pattern that is linked to increased risk for illness, injury, disability or death. The behaviour is usually that of the people whose health is in question, but often, as with sickle cell prevention, it may be the behaviour of those in control of resources, rewards or opportunities, such as parents, peers, teachers, employers, community leaders and health professionals (Green, Kreuter, Deeds and Partridge, 1980; Milner and Desforges 1982; Wethers, 1984; Wethers and Whitten, 1985). Whether a health education programme is operating at the primary, secondary or tertiary stage of prevention, it may accurately be seen as an intervention, the purpose of which is to short-circuit illness or to enhance the quality of life through change or development of health related behaviour. It is expected that such health

behaviour must be voluntary except in cases where the health of others is threatened (Green et al., 1980; Grover, 1980). Voluntary submission of self for haemoglobin genotype screening and readiness to abide by test are the primary preventive health behaviours expected from people who are at risk of having sickle cell offspring.

Generally, screening provides the individual with genetic information of potential value which could assist him in making informed decisions about future reproduction. This benefit of screening notwithstanding, submission of self for the screening test has not received widespread acceptance, particularly in the developing world. Among the many factors identified for this trend are doubts that accurate diagnosis is possible. There is also the perception that without an effective treatment, early diagnosis would not decrease morbidity and mortality. Other reasons are uncertainty about whom to test and questions about obligations to those identified as carriers (Doris, 1989).

In Nigeria, as in most developing countries, adherence to traditional ways of verifying family histories prior to acceptance of marriage proposals by parents, which places emphasis on the family's reproductive history and absence of mental illness, militates against screening through blood test, a concept that is alien to the culture (Arasomwan, 1987; Leigh, 1987). There is

also the belief that marriage is too divine for entrants into the institution to base their decision on some scientific laboratory results (Leigh, 1987; Karanja, 1988). It is also alleged that modernization and education have yet to make marriage an individual rather than family affair. Thus ignorance about the benefit of screening among the broader set of family decision makers disadvantageously affects the utilization of screening services (Pearson, 1974; Powars, 1975; Rogers, 1978; Grover, 1980; 1983; Gaston, Verter and Woods, 1986).

Furthermore, superstitious beliefs about the disease and stigmatization of victims are major issues adversely affecting the willingness of the public to embrace screening. Other constraints include barriers to screening arising from undersupply of preventive counselling services in clinics and hospitals (Olatawura, 1976; Kenen et al 1978; Falusi, 1989; Dada, 1989; Akinyanju, 1990; Anionwu, 1990).

Various behavioural scientists have attempted to give explanations for the factors that militate against the acceptance and utilization of sickle cell screening programmes. But perhaps the most formidable explanations were those drawn from the Health Belief Model.

Health Belief Model

The Health Belief Model proposed by Rosenstock (1966) was

earlier developed in the 1950s to explain preventive health behaviour such as voluntary seeking of immunization services and screening for sickle cell disease (when not mandatory). The model (Figure 1) assumes that the perception and knowledge of people about a particular subject are critical determinants of their health-related behaviour. The model holds that when cues to actions such as relationships with a sickler, mass media campaigns, lectures at school, information from friends and colleagues are present, then the likelihood of undertaking a recommended preventive health measure, such as seeking screening, are dependent on the following four sets of related variables (Rosenstock, 1974).

1. the individual's view of his own vulnerability to the illness or disease condition.
2. his belief about the severity of the illness (this may be defined in term of physical harm or interference with social functioning);
3. the person's perception of the benefits associated with recommended actions to reduce the level of threat or vulnerability; and
4. his evaluation of potential barriers associated with the proposed action (this may be physical, psychological or financial) compared to potential benefits.

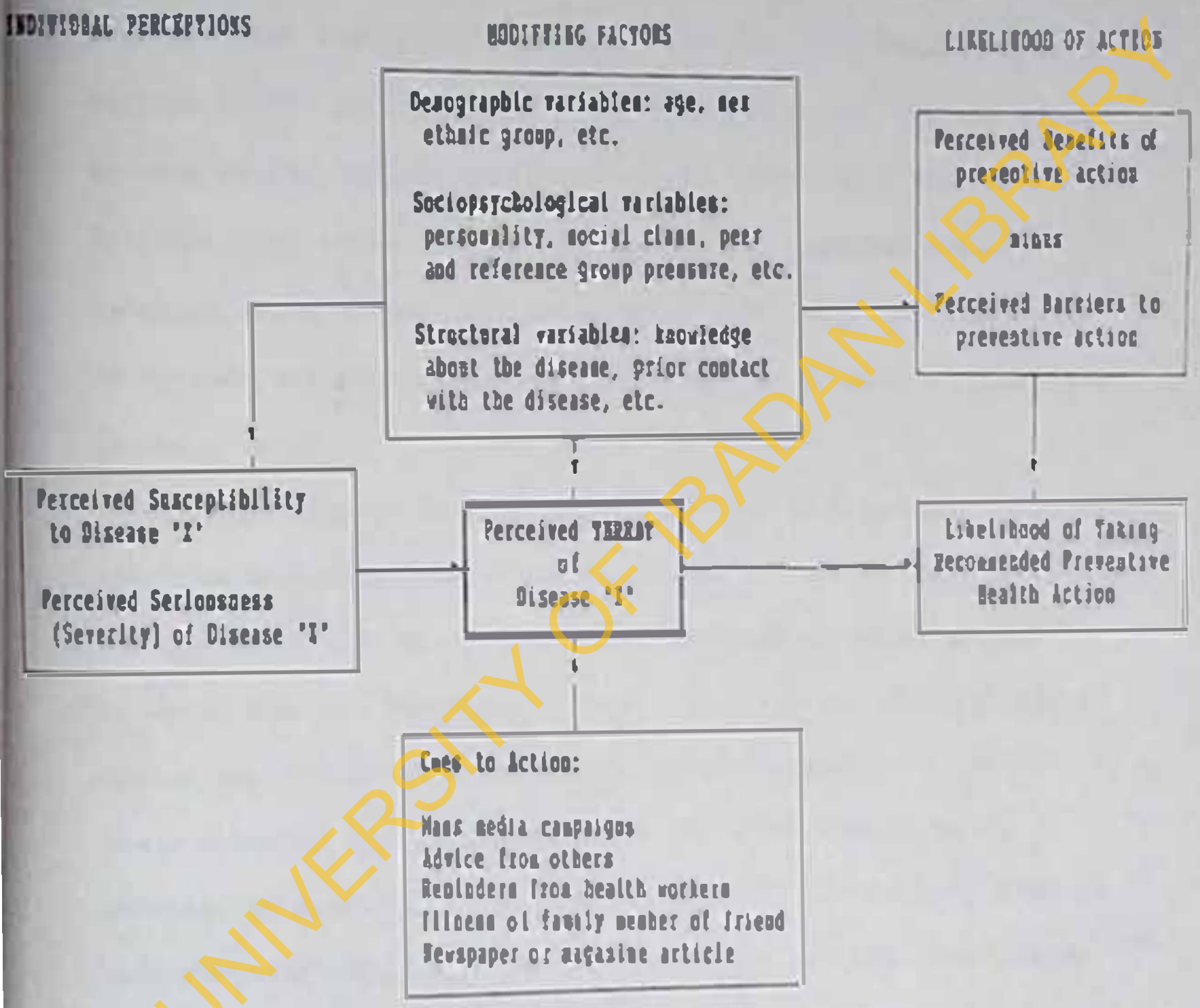


FIGURE 1: The Health Belief Model

Thus, the probability of taking preventive action is a function of the level of perceived threat which is determined by beliefs about susceptibility and severity of the disease. Diaz-Barrios (1989) in a study investigating follow-up response among mothers of 240 infants with sickle cell traits in black and Hispanic population of South Bronx, U.S.A., reported higher response among mothers who anticipated that their children would be victims and who considered the disease as capable of resulting in child death.

Other determinants of human behaviour were perception about benefits derivable from proposed actions to counter the threat and the estimated cost or inconvenience involved in pursuing the proposed action. Kenen and Schmidt (1978) in an investigation of social implication of heterozygote genetic screening in Accra, Ghana observed that respondents who perceived the screening exercise as beneficial complained less about the cost and time of screening and were more likely to persuade relatives and friends to undertake the test.

The relationship between the Health Belief Model and the sickle cell screening phenomenon is evident in Figure 2, which is derived from the findings of several studies described below. In this figure, cues to action that are likely to influence the willingness to seek screening include experiences with sickle cell

patients in the family and information about sickle cell disease from class at school, media, friends, relatives and health workers. Pass, Gauvreau and Schedlhauer (1986) established the relationship between familiarity with persons having sickle cell disease and willingness to seek screening at a new born screening programme for sickle cell disease in New York. There was greater response among mothers who had a sickler in prevlous pregnancies than among those who, though they were at risk, had never had a sickler in previous pregnancies.

Percelved benefit of the action is another important determinant of likelihood to undertaking screening. Sebanjo and Kalu (1989) reported greater clinic attendance among women who believed that early detection of cervical cancer could reduce the adverse effect of the disease. In the report 89% of the 26 patients who believed that early detection of cervical cancer could reduce the complications of the disease attended the screening centre regularly, while only 13% of the 17 patients who believed that the condition once diagnosed could not be ameliorated attended regularly.

Another dimension of the issue of perceived benefit of screening is the fact that most adult populations do not consider screening beneficial to themselves, since information derived from screening and the preventive action expected thereafter was more

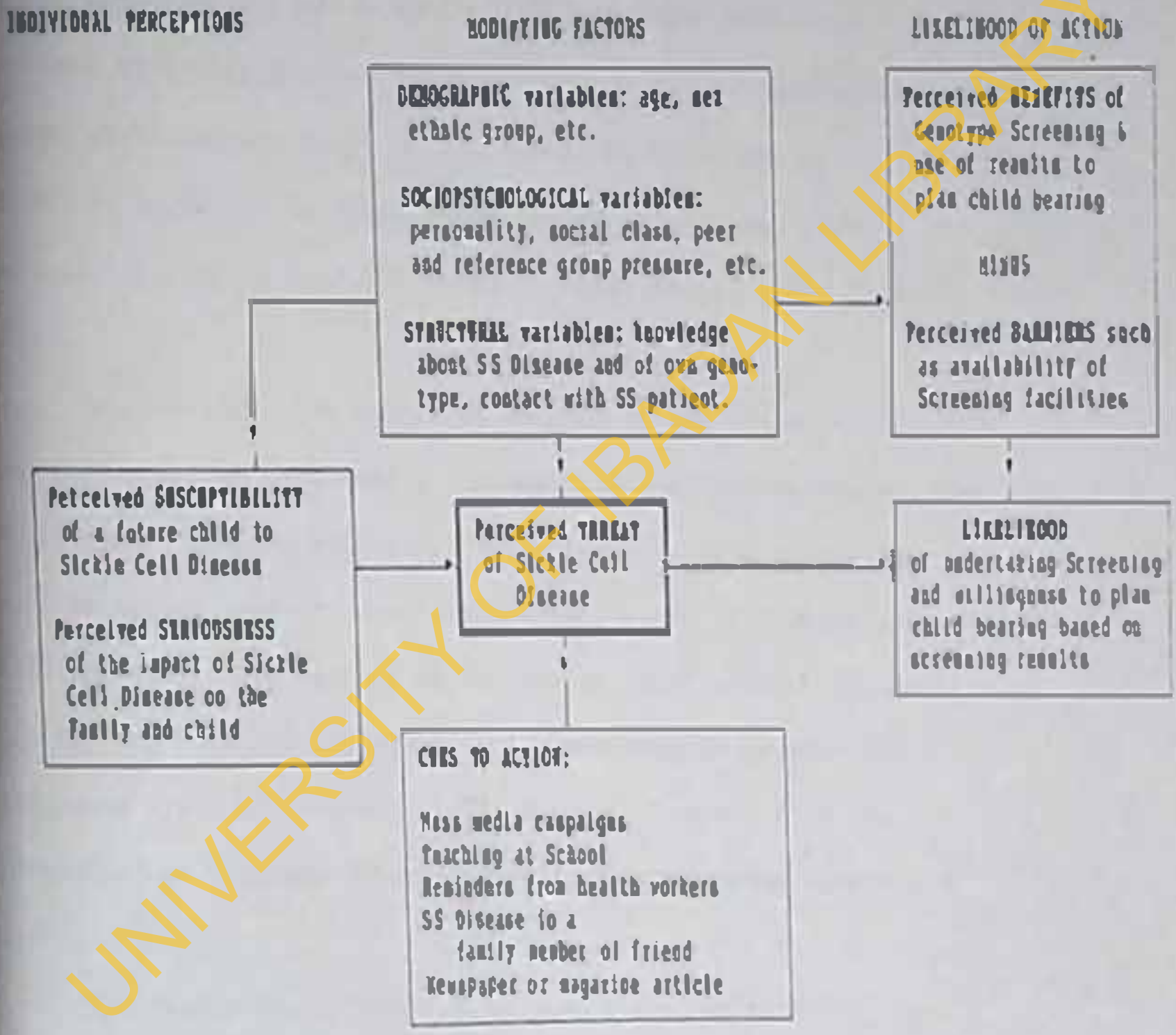


FIGURE 2: Application of the Health Belief Model to Sickle Cell Prevention Activities

for the benefit of the unborn child. Scott (1976) observed that the greater the degree of threat to the personal health of the individual, the more likely it is that he would comply with recommended preventive health action. The situation is made worse by the erroneous belief by adults that attaining adulthood subsumes that they are not sicklers because of the superstitious belief that sicklers taunt their parents by dying young and therefore could not be found among adults (Olatawura, 1976; Calloway and Harrison, 1977; Fleming, 1979; Kaine, 1983; Ebomoyi, 1986).

Incidentally, the perceived benefit of the proposed preventive action, pre-marital haemoglobin genotype screening, has implications for the individual's readiness to make sacrifices such as making time and money available to undertake screening and willingness to face reality by abiding by test result. It also has bearing with the individual's preparedness to protect his offspring from the incurable affliction and himself from the psychological problems associated with the parenthood (Kenen and Schmidt, 1978).

The Health Belief Model also recognizes the fact that an individual's perception about proposed preventive action and barriers hindering compliance are functions of another set of factors called modifiers. Modifying factors in the sickle cell

prevention phenomenon include such variables as marital status, course studied, knowledge about the disease, sex and religion (Ojuyenum, 1980; Ebomoyi, 1986; Leigh 1987). Also age, according to Rogers and Shoemaker (1971) has a modifying effect on people's perception. They postulated that middle aged people and older adults, rather than young adults, are likely to perceive social problems, such as coping with a child with sickle cell disease, more seriously because of their experiences. For the same reason married persons are more likely to perceive the condition more seriously since they may have began facing the realities of parenthood.

At another level, the individuals' perceived seriousness of the disease and perceived susceptibility of self and own children to the trait and disease respectively also influence the decision to seek screening and readiness to abide by the test result which is a higher level of action in the Health Belief framework. It should be noted however that the modifying factors earlier mentioned also determine an individual's perception about the condition. Marital status seems to have a positive influence on the perceived seriousness of the disease as more married than unmarried respondents perceived the disease as a serious problem in the findings of Bamisaiye, Bakare and Olatawura (1974).

One factor that is accountable for the wrong perception of the seriousness of the sickle cell problem in Nigeria is the low level of awareness about the condition among the general public, including the elites (Borrofice, 1979; 1981; Anionwu and Jibril, 1986). Yet it has been reported that in Nigeria, individuals with sickle cell trait make up about 25% of the population and those with sickle cell disease are about 3%, the largest recorded in a single country in the world (Santer, 1976; Fleming, 1979; Kaine, 1983; Akinyanju, 1985). Despite this high sickling rate among the population, a majority of parents and would-be parents continue to perceive their future children as not susceptible to the disease (Olatawura, 1976; Fleming 1979; Borrofice 1981; Falusi 1990).

However, Lesi (1985), while illustrating genotypic distribution amongst families with AS/AS union, observed that the natural law of inheritance reckons that the proportion of genotypes inherited will be 1:2:1 for AA (normal), AS (trait) and SS (sickler) respectively. But he was quick to add that this is not always the case as an analysis carried out in the Lagos University Teaching Hospital showed that only 27% of such families have the so called perfect distribution. It was on the basis of the above that he warned that genetic counsellors must explain to AS/AS parents that the chance of producing a sickler is one in four with <u>each</u> pregnancy.

While Okadiran (1989) asserted that people of low socio-economic status have low level of knowledge of topical and social issues, Cohen (1979) decried that even among practicing nurses and senior nursing students, the percent with adequate knowledge on Genetic conditions was low. He therefore recommended efforts geared towards improving the basic knowledge of all individuals, including health professionals, in all aspects of commonly encountered genetic disorders for the purpose of promoting positive behaviour. In so doing, emphasis should be laid on correcting wrong beliefs, strengthening personal knowledge and addressing the structural and psychological factors that enable an individual to undertake recommended preventive measures.

Summary

In the foregoing chapter, the major issues relating to sickle cell disease and its prevention have been highlighted. First, the nature and extent of the disease with particular emphasis on the history, cause and distribution of the sickle cell gene in populations was enumerated. This was followed by a discussion on the problems faced by families having sickle cell children. Here, issues included the socio-economic and emotional "cost" of morbidity and the strain placed on families in the course of managing sickle cell children.

Screening and counselling services in relation to sickle

cell disease prevention were then considered. In this section, types, technologies, availability and benefits of screening and the objectives of genetic counselling were elaborated. Finally, the chapter concluded with a consideration of preventive health behaviours using the Health Belief Model. The relationship between perceived seriousness and susceptibility, modified by social and structural factors, and the likelihood of undertaking screening, the proposed preventive measure, were discussed. Also, considered were the influences of cues to action and perceived benefits and barriers on perceived disease threat and subsequently, on willingness to undertake the recommended preventive action. These concepts aided in the development of a study instrument as described in the next chapter.

CHAPTER THREE

METHODOLOGY

As mentioned earlier, this research could be seen as a baseline or health education diagnosis of NYSC members' knowledge, attitudes and practices towards sickle cell disease and preventive measures. This chapter describes the methods by which the diagnostic information was obtained. The study population and sampling methods are described. Development, pretesting, administration, validity and reliability of the study instrument are presented. The chapter rounds up with consideration of data analysis methods and study limitations.

DESIGN AND SCOPE

This study is an exploratory, descriptive survey aimed at identifying and documenting the knowledge and perception of Oyo State 1990/91 service year corps members towards sickle cell disease and pre-marital haemoglobin genotype screening. The investigator attempted to identify characteristics of the study population that were associated with their knowledge and perception about sickle cell disease and their willingness to undertake pre-marital haemoglobin genotype screening as a Preventive measure.

Specifically, relationships between demographic variables like sex, religion, marital status, course studied, and presence of known sickle cell patient in respondents' immediate family or environment were tested against respondents' knowledge and perceptions about the disease. The study examined associations between enumerated demographic variables and respondents' willingness to undertake the recommended preventive measure (i.e. genotype screening).

The study was limited to the 1990/91 National Youth Service Corps in Oyo State. The time limit was determined by the length of the researcher's period of study leave. The focus on one state was predicated on the NYSC policy of distributing corps members throughout the country without regard to state of origin. NYSC members in any state of the Federation should share common characteristics. They are drawn from all ethnic groups of the country; are between ages 19-30 years; are all Nigerians; are graduates of either foreign of local polytechnics and universities; are males and females; are mostly unmarried; belong to all common religions; and hold various professional and non-professional certificates. Table 3 shows the distribution of the study population according to state of origin.

TABLE 3

State of Origin of October 1990 NYSC Members In Oyo State

State	Frequency	Percent
Anambra	142	17.0
Akwa Ibom	30	3.6
Bauchi	10	1.2
Bendel	91	10.8
Benue	48	5.7
Borno	13	1.5
Cross River	24	2.8
Federal Territory	3	0.4
Gongola	24	2.8
Imo	143	17.0
Kaduna	10	1.2
Kano	18	2.1
Katsina	5	0.6
Kwara	33	3.9
Lagos	40	4.8
Niger	21	2.5
Ogun	46	5.5
Ondo	50	6.0
Oyo	1	0.1
Plateau	44	5.2
Rivers	29	3.5
Sokoto	15	1.8
TOTAL	840	100

Source: NYSC 1990 33rd bi-annual Conference, Official Report

STUDY VARIABLES

The Health Belief Model (Rosenstock, 1974) was used as the conceptual framework for understanding young people's willingness to undertake pre-marital haemoglobin genotype screening. The operationalization of the variables within this model are described below.

In general, "disease threat" in the case of preventing sickle cell disease must be considered in a different context than the application of the model to an issue like breast cancer screening. In the latter, the individual takes action to protect herself. In the former, the individual takes action to protect his/her children's health and by implication, the well being of his/her future family. In this case, the seriousness must be perceived in relation to how sickle cell disease might disrupt family life of which the individual is an integral part, while perceived susceptibility relates to an unborn child, not the individual making the decision about screening.

"Perceived susceptibility" therefore, is determined in this study by asking the respondent how likely he or she thinks that he/she is capable of giving birth to a child with sickle cell disease. "Perceived seriousness" is elicited by asking respondents to indicate their perception of sickle cell disease with regards to the seriousness of the problem. Options range

between "Not at all serious" to " a very serious problem".

Important modifying factors in this study include sex, religion, marital status, course studied and knowledge. "Knowledge," a major modifying factor, is determined by asking the respondents to state in clear terms, the definition, transmission process, symptoms and modes of prevention of sickle cell disease. Answers cumulatively form a knowledge score.

"Cues to action" are operationally defined to include experience with sickle cell patients in the respondents' own family or immediate environment and information about sickle cell disease received through media, school and friends.

"Perceived benefits" are elicited by asking the respondents to state why it is necessary for people to undertake screening for haemoglobin genotype. "Perceived barriers" are deduced by asking respondents to list the imagined obstacles preventing people from undertaking haemoglobin genotype test.

"Likelihood of action" is defined either as actual completion of screening or intention to undertake it. This is the major dependent variable of the study.

OBJECTIVES OF THE STUDY

1. To document the knowledge and perception of NYSC members about cause, problems and prevention of sickle cell disease.
2. To identify factors associated with the existing level of

knowledge and perception.

3. To determine NYSC members willingness to undertake pre-marital haemoglobin genotype screening.

4. To identify reasons for and factors associated with their willingness to be screened.

5. To draw implications for NYSC members' orientation and for community health education from the findings.

STUDY HYPOTHESES

Hypotheses in this study refer to possible associations between independent and dependent variables. All hypotheses are framed as null hypotheses.

1. There is no significant relationship between sex of respondents and NYSC members' knowledge and perception of sickle cell disease and willingness to undertake haemoglobin genotype screening.

2. There is no significant relationship between religion of respondents and NYSC members' knowledge and perception of sickle cell disease and willingness to undertake haemoglobin genotype screening.

3. There is no significant relationship between marital status and NYSC members' knowledge and perception of sickle cell disease and willingness to undertake haemoglobin genotype screening.

4. There is no significant relationship between course studied and NYSC members' knowledge and perception of sickle cell disease and willingness to undertake haemoglobin screening.

5. There is no significant relationship between familiarity with persons having sickle disease and NYSC members' knowledge and perception of sickle cell disease and willingness to undertake haemoglobin genotype screening.

STUDY POPULATION

The study population consisted of all NYSC members posted to Oyo State in October 1990. The contribution of the then 21 states and the Federal Capital Territory to the NYSC member population in Oyo State during the service year is shown in Table 3. Also, the distribution of NYSC members among the then 42 LGAs in the state, is seen in Table 4.

Sampling

The distribution of the 840 NYSC members around the state, was such that four hundred and twenty, representing 50 per cent, were deployed within Ibadan Municipal Government (IMG). The rest were posted to the remaining former 41 Local Government Areas (LGAs). Thus, using location as criterion, the researcher stratified the study population into I.M.G.and other L.G.As. Stratified random sampling was used to choose respondents from both the municipality and the LGAs.

TABLE 4

Distribution of October 1990 NYSC Members in All LGAs of Oyo State

LGA	No.	%
Ibadan Municipal	420	50.0
Afijio	15	1.8
Aiyedade	9	1.1
Aiyedire	7	0.8
Akinyele	15	1.8
Atakumosa	6	0.7
Boripe	8	0.9
Ede	11	1.3
Egbeda	7	0.8
Egbedore	7	0.8
Ejigbo	8	0.9
Ibarapa	7	0.8
Ido	6	0.7
Ife-Central	10	1.2
Ifedapo	5	0.6
Ifelodun	8	0.9
Ifeloju	4	0.5
Ife-North	7	0.8
Ife-South	8	0.9
Ila	7	0.8
Ilesha	14	1.6
Irepo	7	0.8
Irepodun	10	1.2
Irewole	9	1.1
Iseyin	13	1.5
Iwo	13	1.5
Kajola	7	0.8
Lagelu	8	0.9
Obokun	10	1.2
Odo-Otin	9	1.1
Ogbomoso	31	3.7
Ogo-Oluwa	9	1.1
Ola-Oluwa	9	1.1
Olorunda	9	1.1
Oluyole	9	1.1
Ona-Ara	8	0.9
Orelope	9	1.1
Oriade	9	1.1
Orire	8	0.9
Osogbo	30	3.6
Oyo	25	2.9
Surulere	9	1.1
TOTAL	840	100

In locating NMC respondents, their organization into five community development groups according to day of the week was utilized. Two days (that is two groups) of five community development assignment days were chosen by ballot (Monday and Thursday). All NYSC members who were scheduled to participate in the community development exercise on the chosen days were included as respondents.

For the rest of the state, one-third (i.e. fourteen) of the 41 Local Government Areas were selected through balloting, and all NYSC members serving in the chosen LGAs were included in the sample.

THE STUDY INSTRUMENT

A 34 item self-administered questionnaire made up of sections A and B, served as the study instrument. Section A, comprising questions one to seven, sought respondents' demographic characteristics such as age, sex, religion, marital status, ethnic group, type of institution attended and course studied. Questions eight to 34 comprised section B. This part commenced with questions eight to twelve, which measured respondents' awareness about sickle cell disease and sources from where information about the disease were received. Questions 13 to 22, with the exception of question 18, measured respondents' knowledge of sickle cell disease against a maximum score of 17 points and a minimum score

of zero. Question 18, measured respondents' perceived "Disease Threat" in terms of impact of the disease on the family.

Questions 23 to 25, sought to identify respondents who have already undertaken haemoglobin genotype test and reasons for undertaking the test. The willingness of respondents yet to undertake the test to seek it, was measured by question 26, while questions 28 to 30 identified the various levels of familiarity of respondents with persons having the sickle cell disease. Question 32b measured respondents willingness to abide by sickle cell test results, while question 34 sought to identify respondents' perceived barriers to undertaking the test.

ADMINISTRATION OF THE INSTRUMENT

Official permission was sought and obtained from the Oyo State NYSC Authority to train and make use of the schemes' 14 selected LGA Liaison Officers in the administration of the questionnaire. The fourteen selected LGA Liaison staff assembled at the conference room of the State NYSC office to receive orientation from the researcher on how to administer the instrument with minimal bias such as may be caused through sharing of information among respondents. The orientation exercise lasted six hours during which the aims of the research, the structure and content of the instrument, time, mode of administration, dates and locations of administration and expected roles and conduct of the

liaison staff were discussed.

As the researcher was solely responsible for the administration of the instrument among IMG NYSC members, a pre-administration visitation was made to the Community Development Centres where NYSC members work on Mondays and Thursdays . These were the Motherless Babies Home, Yemetu; the Femi Johnson Insurance House, Oke Ado; the Mapo Hall, Mapo; and at the NYSC Free Consultative Legal and Health Centres, Secretariat, Ibadan. Apart from familiarizing himself with the centres and their managers, the researcher also explained the purpose of the instrument to the centres' operators during the visits.

Pre-testing of the instrument was carried out among fifteen NYSC members of Akinyele LGA who were attending NYSC members LGA meeting at the LGA Secretariat, Moniya. Akinyele LGA had been excluded from the sample by balloting. After the pre-testing, a 45-minute meeting was held with the respondents to evaluate the simplicity and specificity of the questions. The result of the evaluation exercise culminated in the final instrument which was then retested among twenty 1989/90 passing out NYSC members during their final clearance at the states' NYSC Secretariat, Old Ife Road, Ibadan.

Thereafter, the self-administered questionnaire in English Language was completed by respondents under the supervision of the

researcher when NYSC members gathered for Community Development activities in IMG, and the fourteen NYSC liaison staff in the other LGAs when members reported to claim their monthly stipend. The proportions of IMG NYSC members and those of the other 14 LGAs in the sample are detailed in Chapter Four.

DATA ANALYSIS

The data once collected was manually sorted, edited and coded. Frequency tables and diagrams were used to describe the variables. Analysis of variance (F test) was conducted for knowledge scores while the chi-squared (X^2) test was used to examine potential associations between willingness to undertake genotype screening and independent variables.

VALIDITY AND RELIABILITY

The content validity of the instrument was verified both from literature and by members of Faculty of Clinical Sciences and Dentistry of the University of Ibadan who attended the seminar where the initial research protocol was presented. This was further established by identifying the proportion of responses to specific open ended questions at both pre-test and re-test stages that gave the impression that certain questions were ambiguous. Questions nine and twelve, which were originally open ended, had to be reconstructed as closed ended as a result of such observations before the final administration of the instrument.

For reliability, the researcher ensured that instructions on the questionnaire were simple and clear such that could be understood consistently by all respondents. Furthermore, the researcher assured both the pre-testing and re-testing of the instruments were carried out among NYSC members in LGAs and service groups other than those included in the study by ballot.

LIMITATIONS

The major limitation in this study is the inability of the researcher to provide haemoglobin genotype testing facilities to verify respondents' claimed willingness to undertake the test. The result is that the study relied solely on reported willingness to undertake test as expressed by respondents on the questionnaire.

Also it is possible that some of the respondents may have undertaken the haemoglobin genotype test much earlier in their lives and did not remember the event. Again the information on those already screened relies on their reports alone. This and the memory factor place limitations on the reported distribution of blood types reported in the next chapter.

CHAPTER FOUR

RESULTS

Two hundred and Ninety-nine questionnaires out of three hundred and eleven distributed to respondents were returned. Among these, eight were not fully completed and were thus discarded from data analysis, yielding a final sample size of 291. Distribution of respondents among the chosen LGAs is seen in Table 5.

DEMOGRAPHIC CHARACTERISTICS OF RESPONDENTS

The distribution of the respondents with regard to age, sex, marital status, religion, ethnic group, type of higher institution attended and course studied are described in this first section. The largest number of respondents (43.2%) were in the age bracket 25-27 years. This was followed by 29.5% who were between 22-24 years old, while 19.5% were in the age group 28-30 years. Only 7.2% were in the age range 19-21 years (see Figure 3). Most of the respondents (62.5%) were males, while the remainder (37.5%) were females. With regards to marital status, a larger percentage (79%) of the respondents were single, while 17% were either engaged or introduced to their future partners. A few (4%) were already married.

With respect to religion, majority of the respondents (78.4%)

TABLE 5

DISTRIBUTION OF RESPONDENTS AMONG THE CHOSEN LOCAL GOVERNMENT AREAS

Local Government	Number	Percent
Ibadan Municipal	150*	51.6
Alyedade	9	3.1
Ede	11	3.8
Ibarapa	7	2.4
Ido	6	2.1
Ifelodun	8	2.7
Ife North	7	2.4
Ife South	8	2.7
Ilesha	14	3.8
Irepo	7	2.4
Lagelu	8	2.7
Odo-Otin	9	3.1
Ogbomosho	31	10.8
Ona-Ara	8	2.7
Orire	8	2.7
TOTAL	291	100

* 150 of 420 NYSC Members in Ibadan Municipal were selected, while all were chosen in the remaining LGAs.

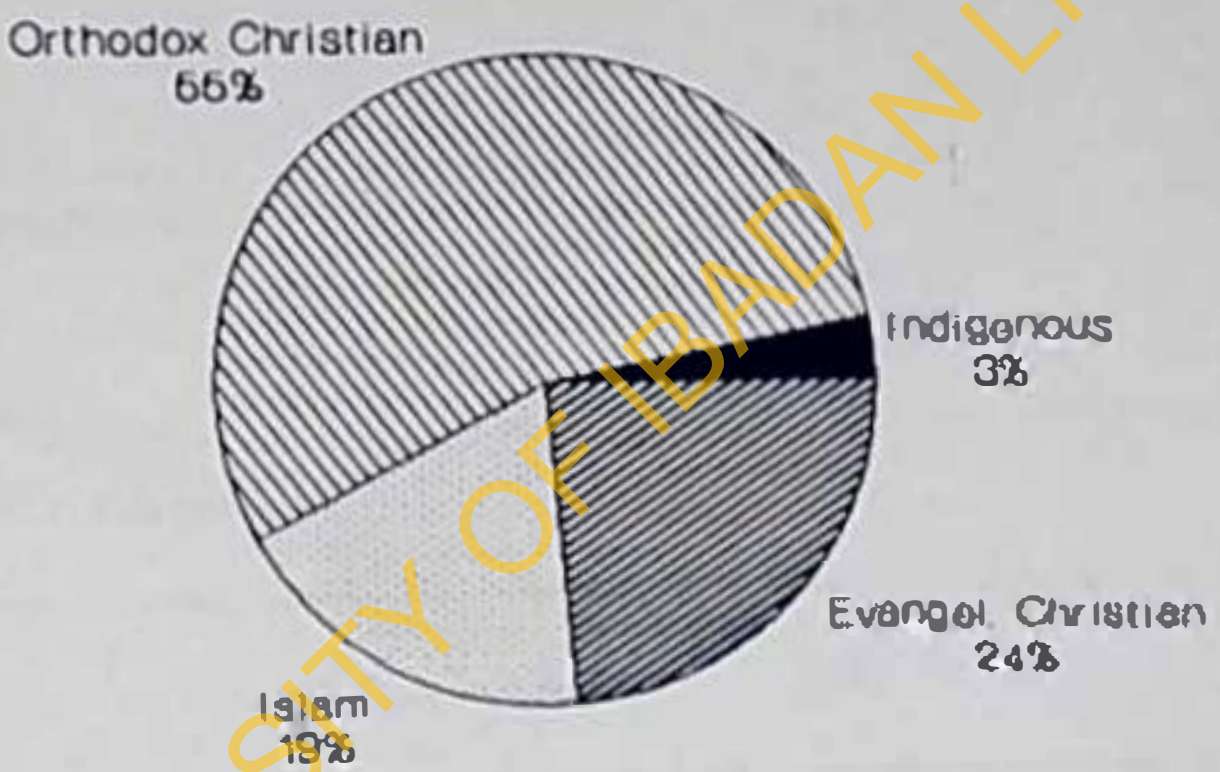
FIGURE 3: Age Distribution of Respondents

were Christians. Of these, orthodox church worshippers, made up of Baptist, Catholic, Anglican and Methodist, were 54.6%. The other Christians (23.8%) belonged to syncretic and evangelical churches made up of Aladura, Celestial, Apostolic, Seraphim and Cherubim and Brotherhood of the Cross and Star. Of the remaining respondents 18.5% were Muslims, while only 3.1% were indigenous religion worshippers (Figure 4).

Table 6, shows that more than a third (34%) of the respondents were Ibo. About one-fifth (21%) were Yoruba. Other groups included Hausa/Fulani (10%); other northerners (14%); Efik and other Easterners (9%), and Bendelites (12%).

Most of the respondents (74%) attended Universities, while about a quarter (26%) completed their higher education at polytechnics. In respect of course studied, majority (45%) studied either Social Sciences or Arts. Eighty-Five (29.2%) read either physical sciences, made up of physics, chemistry, mathematics, computer science, statistics and engineering. Some forty-seven (19.6%) majored in either biological (botany, zoology, biochemistry, microbiology) or agricultural sciences. Only 18 (6.2%) studied health sciences made up of medicine and surgery, nursing, pharmacy, physiology, anatomy, medical microbiology and laboratory technology (see Figure 5).

Religion of Respondents



N = 291

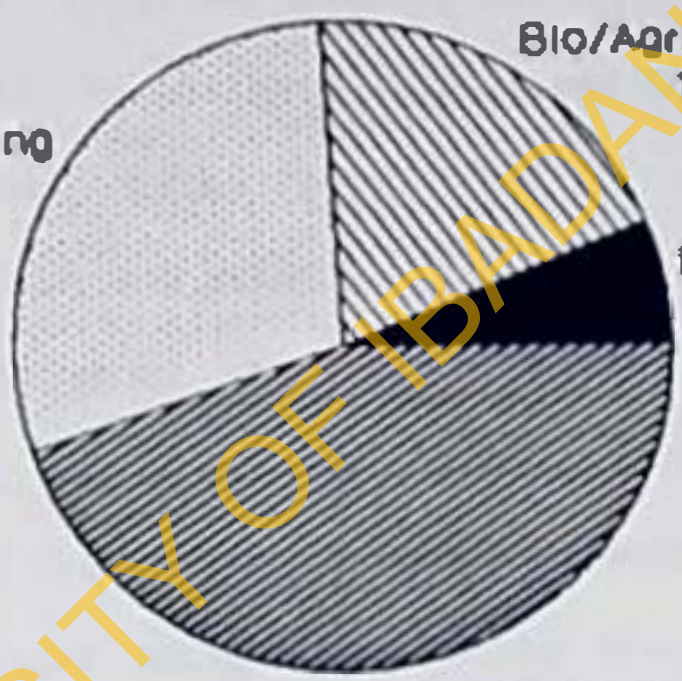
FIGURE 4: Distribution of Respondents According to Religion

TABLE 6

Ethnic Distribution of Respondents

Ethnic Group	Frequency	Percent
Ibo	99	34
Yoruba	61	21
Other Northerners	41	14
Bendalites	33	12
Hausa/Fulani	30	10
Efik & Other Easterners	27	9
TOTAL	291	100

Courses Studied



N = 201

FIGURE 5: Distribution of Respondents According to Course Studied

KNOWLEDGE AND PERCEPTION

The respondents were asked whether they had heard about sickle cell disease before. In response, a majority (84.6%) said they had heard about it, while 15.5% said they had not. In answer to another question which sought when the respondents first heard about it, 62.9% said it was at class in secondary school. Twenty-two (7.5%) said they heard of it first at primary school, 6.9% heard after secondary school but before tertiary school, and finally 7.2% became aware at tertiary school (Table 7).

A further question was asked to show all sources from where respondents had heard about sickle cell disease. The multiple responses showed that 83% of the 246 who had heard of the disease, received information from class teachers at school. Nearly half (47%) heard from friends while 41% heard on television and radio. Some 36% claimed they heard from relatives and 30% read about it from newspapers and magazines. Only 17% received the message from health workers, while 9% received the information at seminars as seen in Figure 6.

Corps members' knowledge about sickle cell disease was based on responses to nine questions (see Appendix 3). A maximum score of 17 points was possible. Questions covered the following topics; definition of sickle cell disease and trait, symptoms, and disease transmission, prevention, treatment and screening issues.

TABLE 7

Distribution of Respondents According to When First Heard About Sickle Cell Disease

When First Heard	Frequency	Percent
During Primary School	22	7.6
During Secondary School	183	62.9
After Secondary School but Before Tertiary	20	6.9
During Tertiary School	21	7.2
Never Heard	45	15.4
TOTAL	291	100

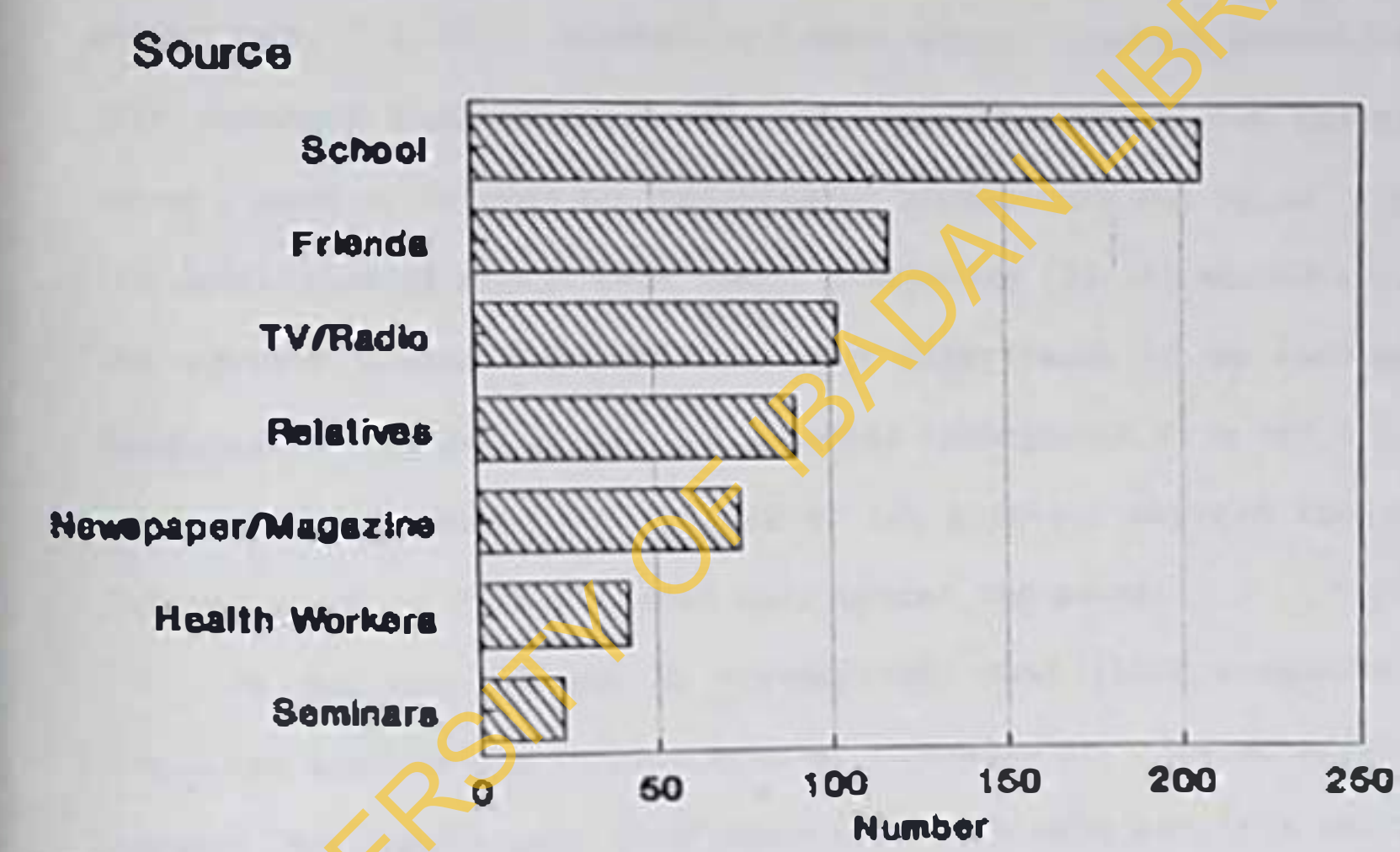


FIGURE 6: Sources From Where Respondents Heard About Sickle Cell Disease

Their scores ranged from zero to 17 points with an average of 8.5 points. The overall mean (5.2) reflects a very low level of knowledge among study population.

Examples of the knowledge problem include the fact that nearly two-thirds (65.6%) could not define the disease. The acceptable answer was, "It is a hereditary haematologic disorder associated with abnormal haemoglobin," which scored the maximum two points. Answers such as "blood" or "hereditary" scored only one point. For the definition of sickle cell trait, a majority (71.1%) scored zero. The correct answer expected was, "the inheritance of an abnormal haemoglobin from one parent and a normal haemoglobin from the other parent," which scored the maximum of two points. Answers such as "haemoglobin" or "inheritance" only scored one point.

On how the disease is transmitted, most (79%) respondents could not mention the correct mode of transmission. The acceptable answer, "by inheritance from parents who are both carriers and/or sicklers," scored the maximum one point. Regarding knowledge of symptoms of the disease, 37.1% scored less than half (three) of the maximum six points allocated. The correct answers expected were fever, painful crisis, infection, dehydration, breathlessness, anaemia (pale appearance), stunted growth, and lack of strength, each of which scored one point.

In respect to knowledge of prevention of the disease, most

respondents (63.5%) said it was not preventable. The acceptable answer was "it was preventable through avoidance of child bearing between two trait carriers or sicklers" which scored the maximum two points. They were also asked "whether there was cure for the disease." In response 43% scored zero. The answer expected was "no cure" which scored the maximum one point. On how to confirm one's sickle cell status, 27.2% did not know what to do to confirm their statue. The expected answer was "by blood test at hospital" which scored the maximum one point.

Also, respondents were asked to state the most appropriate time when healthy adults should confirm their sickle cell status. Thirty six and half percent did not know when. The acceptable answer was "when choosing a marriage partner" which scored the maximum one point. Finally, the respondents were asked to state the benefit of knowing one's sickle cell status. As many as 37.9% did not know the benefit of screening. The acceptable answer was, "it enables individuals to make an informed decision about marriage and reproduction," which scored the maximum one point.

FACTORS ASSOCIATED WITH KNOWLEDGE

In an effort to identify factors associated with existing level of knowledge of the respondents about sickle cell disease, the researcher compared the knowledge score of respondents with the following independent variables: sex, religion, marital status,

course studied, history of screening, perceived seriousness of the disease, perceived susceptibility of own children, familiarity with persons having sickle cell disease and willingness to undertake screening for the disease.

When knowledge score was compared against respondents' sex, a statistically significant difference was found between the mean knowledge score of 5.7 for males and 4.6 for females (Table 8).

Religion and knowledge score were compared by grouping religion into four broad categories - Orthodox Christian, Evangelical and Syncretic Christian, Islam and Indigenous African worshippers. Results revealed that Orthodox Christians had the highest mean knowledge score (6.5) closely followed by adherents to Islamic religion (6.2). Indigenous worshippers came third with a mean score of 3.7, while the Evangelical/Syncretic Christians scored the lowest (1.9). Statistically, these differences are significant as evident in Table 9.

Marital status and knowledge od sickle cell disease were also compared. Respondents who were still single had the highest mean knowledge score of 6.2. Those who were either engaged or already introduced to their future partners had a mean score of 1.9, while others who were married had the lowest (1.4). Table 10 shows that these differences were statistically significant.

TABLE 8

Comparison Between Knowledge Score and Sex

	SEX		
	Male	Female	Total
Mean	5.7	4.6	5.3
Total Score	1,033	504	1,537
Sum of Squares	7,991	3,432	11,423
Number	182	109	291
Standard deviation	3.4	3.2	

F = 6.75, d.f. = 1;289, p < 0.005

TABLE 9

Comparison Between Religion and Knowledge Score

	RELIGION				
	Orthodox Christian	Islam	Indigenous African	Evangelical Christian	Total
Mean	6.5	6.2	3.7	1.9	5.3
Total Score	1,042	332	33	130	1,537
Sum of Squares	8,774	324	2,176	149	11,423
Number	160	54	9	68	291
Standard deviation	3.5	1.6	1.9	1.1	

F = 46.3, d.f. = 3;287, p < 0.0005

TABLE 10

Comparison Between Marital Status and Knowledge Score

	MARITAL STATUS			
	Single	Engaged	Married	Total
Mean	6.2	1.9	1.7	5.3
Total Score	1,429	93	15	1,537
Sum of Squares	11,163	233	27	11,423
Number	231	49	11	291
Standard deviation	3.2	1.1	0.8	

F = 59.4, d.f. = 2:288, p < 0.0001

For comparison of knowledge score and course studied, the various disciplines were grouped into four broad categories - Health Sciences, Biological and Agricultural Sciences, Physical Sciences and Engineering, and Social Sciences and Arts. Results showed that respondents in the health sciences had the highest (12.7) mean knowledge score. This was followed by the score 7.1 for those in Biological and Agricultural Sciences. Others in Physical Sciences and Engineering had a score of 6.7, while the lowest mean knowledge of 2.5 was obtained in the Social Sciences and Arts category. These differences were statistically significant as seen in Table 11.

A comparison was also made between score of respondents and history of screening for the disease. Analysis showed that there was a statistical difference between the mean knowledge score of 8.6 for those who had already undertaken the screening test and 4.7 for those yet to undertake the test as seen in Table 12.

Furthermore, respondents' perceived seriousness of sickle cell disease was compared against their knowledge score. Results showed that those who perceived the disease as a serious problem had the highest mean knowledge score of 6.1 while others who were uncertain about the disease's seriousness and those who perceived it as a simple problem scored 1.7 and 1.4 respectively. These differences were statistically significant as shown in Table 13.

TABLE 11

Comparison Between Course Studied and Knowledge Score

	COURSE STUDIED				
	Health Sciences	Biological & Agric. Sciences	Physical Sci. and Engineer.	Social Sciences & Arts	Total
Mean	12.7	7.1	6.7	2.5	5.3
Total Score	228	404	573	332	1,537
Sum of Squares	2,942	3,378	4,021	1,082	11,423
Number	18	57	85	131	291
Standard deviation	1.8	3.0	1.4	1.4	

F = 231.1, d.f. = 3;287, $p < 0.0001$

TABLE 12

Comparison Between History od Screening and Knowledge Score

	HISTORY OF SCREENING		
	Yes	No	Total
Mean	8.6	4.7	5.3
Total Score	985	552	1,537
Sum of Squares	9,253	2,170	11,423
Number	114	117	291
Standard deviation	2.6	1.6	

F = 6.75, d.f. = 1;289, p < 0.005

TABLE 12

Comparison Between History od Screening and Knowledge Score

	HISTORY OF SCREENING		
	Yes	No	Total
Mean	8.6	4.7	5.3
Total Score	985	552	1,537
Sum of Squares	9.253	2,170	11.423
Number	114	117	291
Standard deviation	2.6	1.6	

F = 6.75, d.f. = 1:289, p < 0.005

TABLE 13

Comparison Between Perceived Seriousness of Sickle Cell Disease and Knowledge Score

	PERCEIVED SERIOUSNESS			
	Very Serious	Uncertain	Not Serious	Total
Mean	6.1	1.7	1.4	5.3
Total Score	1,455	88	44	1,537
Sum of Squares	11,273	70	80	11,423
Number	237	22	32	291
Standard deviation	3.2	0.5	0.8	

F = 57.3, d.f. = 2;288, p < 0.0002

Also, knowledge score was compared with the perceived susceptibility of respondents' own children to the disease. Table 14 shows that there was a statistically significant difference between the mean knowledge score 6.7 for those who say their offspring were likely to be susceptible and 4.7 for others who claimed that their offspring were unlikely.

Familiarity with persons having sickle cell disease and the respondents' knowledge score was another variable that was compared. Results showed that the highest mean knowledge score (11.3) was obtained by those whose brother or sister was a sickler. This was followed by 8.2 for those who had an extended family member who was a sickler. Others who had a sickler as a friend scored 6.2, while those who were only acquainted to sicklers and others who had never met sicklers before scored 4.1 and 1.7 respectively. These differences in mean scores were of statistical significance as shown in Table 15 and Figure 7.

Finally, the knowledge score of those yet to undertake haemoglobin genotype test was compared against their willingness to undertake the test. Table 16 revealed that statistically significant differences exist between the highest mean knowledge score of 4.0 for those who said they were likely to undertake the test and 1.4 for others who were uncertain about undertaking the test and 1.5 for those who said they were unlikely to undertake it.

TABLE 14

Comparison Between Perceived Susceptibility of Own Children and Knowledge Score

	PERCEIVED SUSCEPTIBILITY OF CHILDREN		
	Likely	Unlikely	Total
Mean	6.7	4.7	5.3
Total Score	543	994	1,537
Sum of Squares	4,101	7,322	11,423
Number	81	210	291
Standard deviation	2.4	3.5	

F = 21.3, d.f. = 1;289, P < 0.001

TABLE 15

Comparison Between Familiarity with Person Having Sickle Cell Disease and Knowledge Score

	LEVEL OF FAMILIARITY					
	Brother/ Sister	Extended Family	Friend	Acquainted	None	Total
Mean	11.3	8.2	6.2	4.1	1.7	5.3
Total Score	465	246	363	325	138	1,537
Sum of Squares	5,397	2,042	2,323	1,373	288	11,423
Number	41	30	59	79	82	291
Standard deviation	1.8	0.9	1.2	0.7	0.8	

F = 645.9, d.f. = 4;286, p < 0.00001

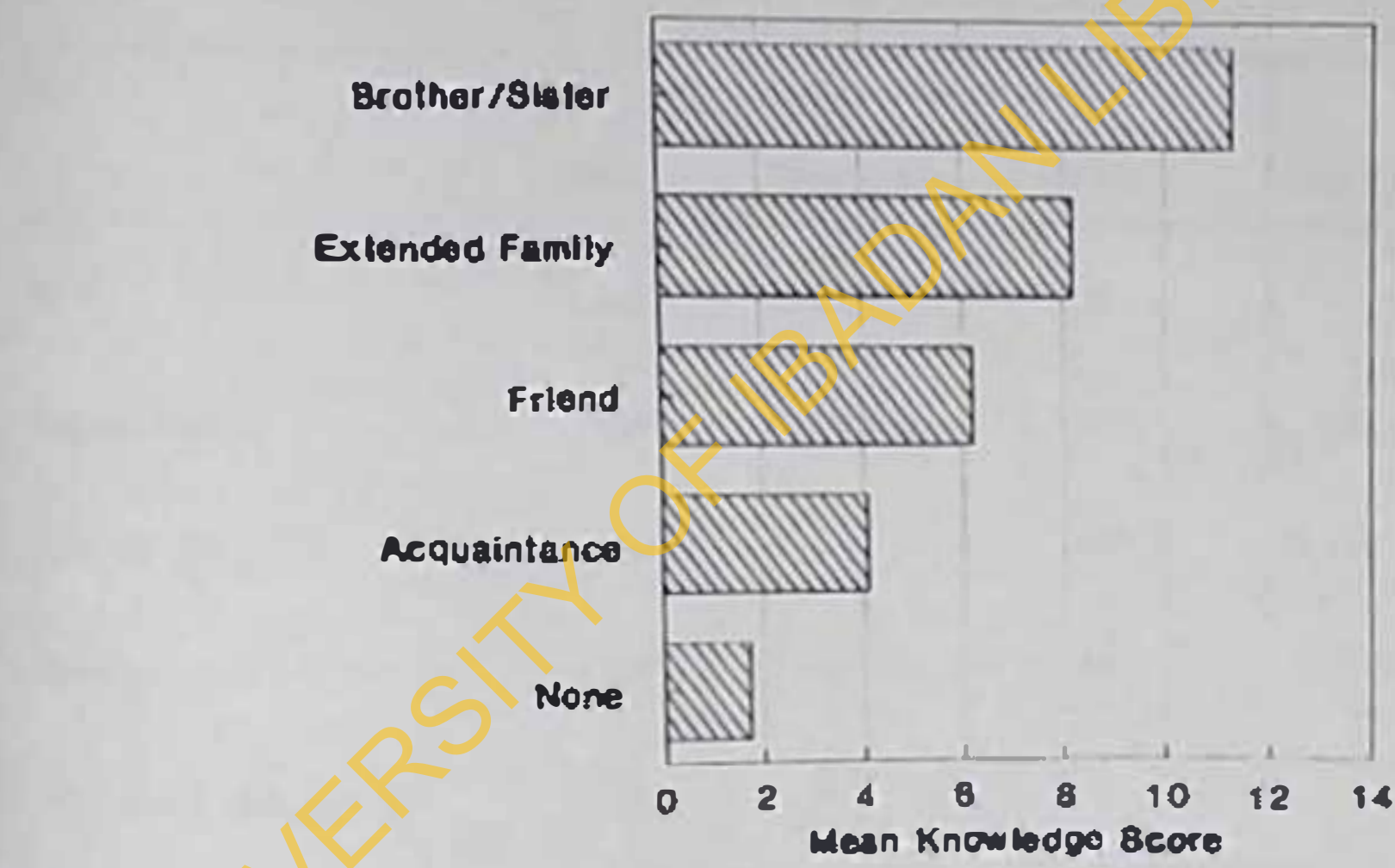


FIGURE 7: Comparison Between Familiarity with a Person Having Sickle Cell Disease and Knowledge Score

TABLE 16

Comparison Between Willingness to Undertake Haemoglobin Genotype Test and Knowledge Score

	WILLINGNESS			
	Likely	Uncertain	Unlikely	Total
Mean	4.0	1.4	1.5	
Total Score	465	21	66	552
Sum of Squares	2,013	37	120	2,170
Number	117	15	45	177*
Standard deviation	1.2	0.7	0.7	

* Those who have not yet taken the test

$F = 112.4$, $d.f. = 2:174$, $p < 0.0002$

HISTORY OF SCREENING

A total of one hundred and fourteen (39.2) respondents claimed they have already undertaken the haemoglobin genotype test prior to this study. According to the Health Belief Model, these represent people who have already taken one step of the recommended preventive action. Of these over half (58.8%) reported that they had normal haemoglobin AA. Thirty-two (28.1%) had AS, while 12.3% had AC. Only one reportedly had the Beta-thal type of sickle cell disease (see Figure 8).

On why these respondents decided to undergo the test, 66.6% said they took the test in fulfillment of requirements for admission to an institution of higher education. Fourteen (12.2%) had it done while they were young by their parents. Ten (8.6%) undertook it in the course of donating blood to patients. Some 4.3% each had taken the test either as part of clinical research or on the basis of mere curiosity. Only four per cent undertook the test to guide their choice of future partners (see Table 17).

In order to distinguish the characteristics of the respondents who had already undertaken haemoglobin genotype test from those who had not, a comparison was made between respondents' sex, marital status, religion, course studied and familiarity with persons having sickle cell disease, and their history of screening.

Reported Genotype

N = 114

FIGURE 8: Reported Genotypic Distribution of Respondents Already Screened

TABLE 17

Distribution of Respondents According to Reasons for Undertaking Hb Genotype Test

Reasons	Frequency	Percent
Admission Requirement	76	66.6
Done by Parents	14	12.2
When Donating Blood	10	8.6
Clinical Research	5	4.3
Curiosity	5	4.3
Guide Choice of Spouse	4	4.0
Total	114*	100

*NOTE: 177 have not yet taken the test

Results revealed that there were no statistical relationships between history of screening and either sex or course studied as shown in Tables 18 and 19. However, religion, marital status, and familiarity with persons having sickle cell disease were found to have significant associations statistically with undertaking the test. Table 20 shows that whereas 45.3% of Orthodox Christians had already undertaken the test, only 33.3% of Muslims and 35.2% of Indigenous religion worshippers had done so. None of the Evangelical/Syncretic Christians had undertaken the test.

Regarding marital status, Table 21 showed that 45% of the single respondents, in contrast to 18.2% of the married and 16.3% of the engaged or introduced had been tested.

With respect to familiarity with persons having sickle cell disease, 73.2% of those with a brother or sister who had sickle cell disease had been tested. This was followed by 43.3% of those with an extended family member and 32.2% with a friend who is a sickler. Also, 31.6% of respondents who are only acquainted with sickle cell patients and 32.9% of those who have never met one were already tested (Table 22 and Figure 9).

TABLE 18

Sex of Respondents Compared with History of Hb Genotype Screening

Already Tested	SEX Male (%)	Female (%)	Total
YES	69 (37.9)	45 (41.3)	114
NO	113 (62.1)	64 (58.7)	177
TOTAL	182	109	291

$X^2_{Yates} = 0.199$, d.f. = 1, $p > 0.65$

TABLE 19

Comparison Between Course Studies and

History of Hb Genotype Screening

Already Tested	COURSE STUDIED (%) Health Sciences	Biological & Agric. Sciences	Physical Sci. and Engineer.	Social Sciences & Arts	Total
YES	8 (44.4)	25 (43.9)	25 (29.4)	56 (42.7)	114
NO	10 (55.6)	32 (56.1)	60 (70.6)	75 (57.3)	177
TOTAL	18	57	85	131	291

X^2 = 4.836, d.f. = 3, p > 0.18

TABLE 20

Religion of Respondents Compared with History of Hb Genotype Screening

Already Tested	RELIGION (%) Orthodox Christian	Islam	Indigenous African	Evangelical Christian	Total
YES	72 (45.3)	19 (35.2)	0 (0)	23 (33.3)	114
NO	87 (54.7)	35 (64.8)	9 (100)	46 (66.7)	177
TOTAL	159	54	9	69	291

χ^2 = 9.635, d.f. = 3, p < 0.022

TABLE 21

Marital Status of Respondents Compared with History of Hb Genotype Screening

Already Tested	MARITAL STATUS (%) Single	Engaged	Married	Total
YES	104 (45.0)	8 (16.3)	2 (18.2)	114
NO	127 (55.0)	41 (83.7)	9 (81.8)	177
TOTAL	231	49	11	291

X^2 = 16.084, d.f. = 2, p < 0.0004

TABLE 22

Familiarity with Persons Having Sickle Cell Disease Compared with History of Hb Genotype Screening

Already Tested	LEVEL OF FAMILIARITY (%) Brother/ Sister	Extended Family	Friend	Acquainted	None	Total
YES	30 (73)	13 (43)	19 (32)	25 (32)	27 (32)	114
NO	11 (27)	17 (57)	40 (68)	54 (68)	55 (68)	177
TOTAL	41	30	59	79	82	291

$X^2 = 24.42$, d.f. = 4, $p < 0.001$

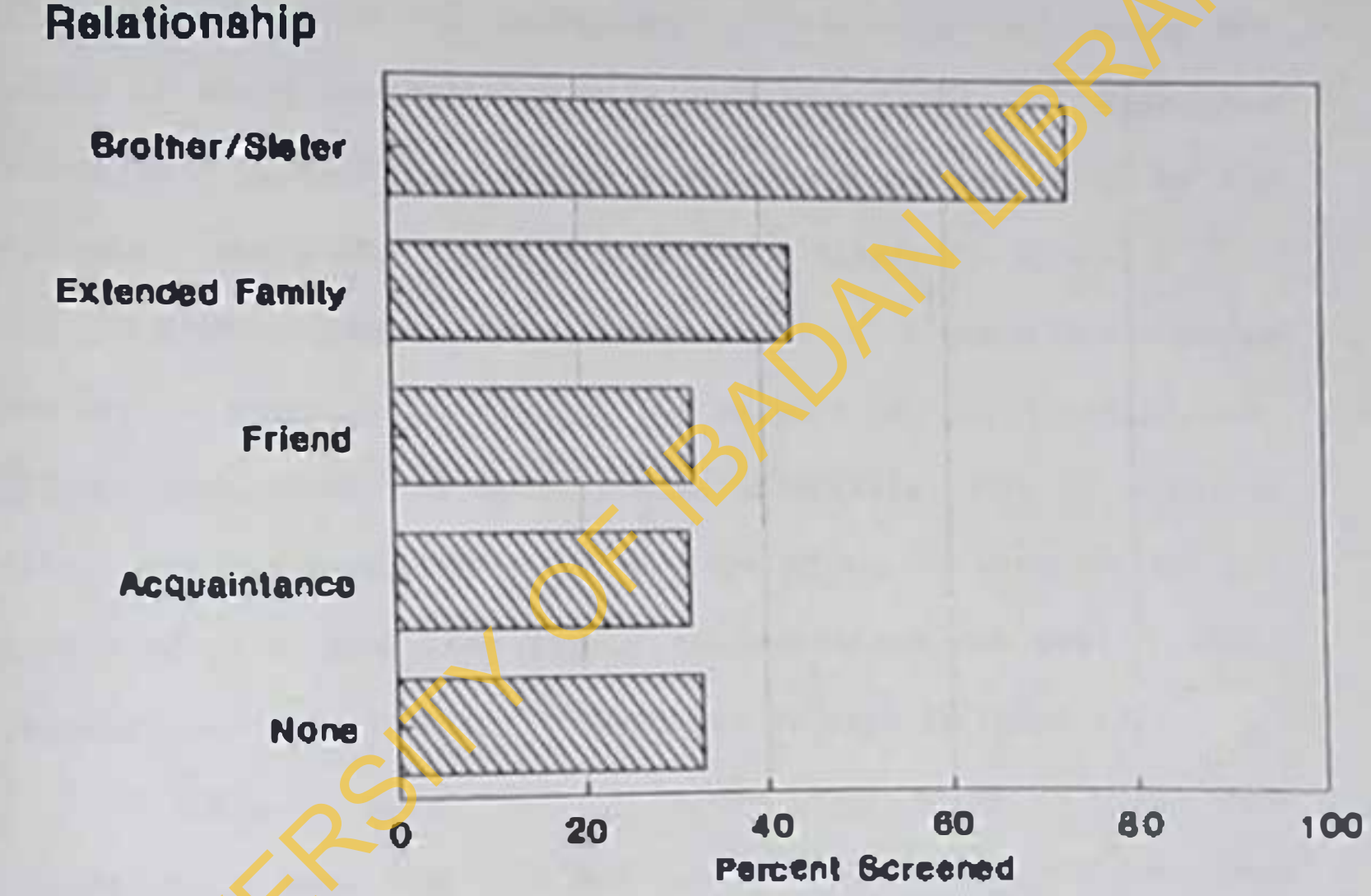


FIGURE 9: Familiarity with Persons Having Sickle Cell Disease and History of Having Been Screened for Hb Genotype

WILLINGNESS TO UNDERTAKE SCREENING

In determining the willingness of the 177 (60.8%) respondents yet to undertake the test to submit themselves for screening, several variables were compared with these Corps members' stated likelihood to undertake screening. First, they were asked how likely it would be that they will undertake the test. Responses showed that a majority (66.1%) were likely to subscribe to the procedure, while 33.9% said they were not likely to do so.

Course studied was then compared with willingness to undertake the test. Analysis showed that 100 percent of health scientists, 76% of biological and agricultural scientists, 60% of physical scientists and engineers and 61 percent of social sciences and art graduates said they were likely to understand the test. These results were statistically significant as seen in Table 23.

Concerning sex, Table 24 shows that males (73.4%) were significantly more likely to say they would undertake the test than females (53.1%). A comparison by religion showed that 65.1% of the Orthodox Christians indicated that they were likely to undertake the screening exercise. This was followed by 71.4% of the Muslims. For the indigenous religion worshippers, 33.3% said they were likely, while 32.6% of the Evangelical and Syncretic Christians indicated willingness to undertake the test. These differences are statistically significant as seen in Table 25.

TABLE 23

Comparison Between Course Studied and Willingness of Untested Respondents to Undertake Hb Genotype Test

Willing to be Tested	COURSE STUDIED (%) Health Sciences	Biological & Agric. Sciences	Physical Sci. and Engineer.	Social Sciences & Arts	Total
Likely	10 (100)	29 (76)	28 (60)	50 (61)	117
Uncertain/ Unlikely	0 (0)	9 (24)	19 (40)	32 (39)	60
TOTAL	10	38	47	82	177
	10	32	60	75	

X^2 = 8.723, d.f. = 3, p < 0.035

TABLE 24

Comparison Between Sex and Willingness of Untested Respondents to Undertake Hb Genotype Test

Willing to be Tested	SEX Male (%)	Female (%)	Total
Likely	83 (73.4)	34 (53.1)	117
Uncertain/ Unlikely	30 (26.6)	30 (46.9)	60
TOTAL	113	64	177

$\chi^2_{Yates} = 6.654$, d.f. = 1 $p < 0.01$

TABLE 25

Religion of Untested Respondents Compared with Willingness to Undertake Hb Genotype Test

Willing to be Tested	RELIGION (%) Orthodox Christian	Islam	Indigenous African	Evangelical Christian	Total
Likely	74 (85.1)	25 (71.4)	3 (33.3)	15 (32.6)	117
Uncertain/ Unlikely	13 (14.9)	10 (28.6)	6 (66.7)	31 (67.4)	60
TOTAL	87	35	9	46	177

χ^2 = 41.736. d.f. = 3. p < 0.00001

Results of comparison between marital status and willingness to be tested revealed that 91.3% of the single respondents indicated likelihood to undertake screening against 2% of either the engaged or married respondents. These difference was of statistical significance as seen in Table 26.

Finally, familiarity with sickler was compared with willingness to undertake the test. Analysis showed that respondents with a brother or sister who was a sickler were more (100%) likely to undertake screening. These were followed by those with friend as sickler (75%) and others who never met sicklers before (70.1%). For those with extended family member as sickler, 58.8% said they were likely to undertake it, while 47.3% of those who have met sicklers but were not related indicated likelihood to undertake screening (see Table 27 and Figure 10).

PERCEPTION ABOUT SICKLE CELL DISEASE

Respondents' perceived seriousness of sickle cell disease was measured through question 18, which sought to find out the perceived impact of the condition on the family. Responses showed the majority (72.1%) agreed that such a child would definitely have a negative impact on family life, while the remaining (27.9%) said this would not. To check the above responses, question 27 was asked which categorized perceived seriousness of the condition as follows: very serious, uncertain, and not serious. Most (81.4%) respondents

TABLE 26

Marital Status of Untested Respondents Compared with Their Willingness to Undertake Hb Genotype Test

Willing to be Tested	MARITAL STATUS (%) Single	Engaged/Married	Total
Likely	116 (91.3)	1 (2.0)	117
Uncertain/ Unlikely	11 (8.7)	49 (98.0)	60
TOTAL	127	50	177

$\chi^2_{Yates} = 123.832$, d.f. = 1, $p < 0.00001$

TABLE 27

Comparison Between Familiarity with Persons Having Sickle Cell Disease and Willingness of Untested Respondents to Undertake Hb Genotype Test

Willing to be Tested	LEVEL OF FAMILIARITY (%) Brother/ Sister	Extended Family	Friend	Acquainted	None	Total
Likely	11 (100)	10 (59)	30 (75)	26 (47)	40 (70)	117
Uncertain /Unlikely	0 (0)	7 (41)	10 (25)	29 (53)	14 (30)	60
TOTAL	11	17	40	55	54	177

χ^2 = 17.690, d.f. = 4. p < 0.0015

FIGURE 10: Familiarity with Persons Having Sickle Cell Disease and Willingness of Untested Respondents to Undertake Hb Genotype Test

said it was a very serious problem. Some 11 percent claimed it was not serious, while 7.6% were uncertain about its seriousness. However, a comparison of responses between those already tested for the Hb genotype and others yet to be tested showed no statistical relationship (see Table 28).

Also, willingness to undertake the Hb Genotype test among the 177 respondents yet untested was compared against their perceived seriousness of the condition. Analysis showed that 78.5% of those who perceived the disease as "very serious" were willing to undertake the test whereas, 58.8% of those who were uncertain about the seriousness of the condition were undecided about taking the test. Furthermore, 75% of those who perceived the disease as "not serious" were willing to undertake the test. These differences are statistically significant as seen in Table 29.

Following the observation of zero and small numbers in some of the cells, respondents who were uncertain were combined with those who claimed that the disease was not serious in the analysis of the relationships between religion, course studied, relationship with known sicklers and perceived seriousness. The results revealed that all three variables were associated with the respondents' perceived seriousness of the condition.

TABLE 28

Perceived Seriousness of Sickle Cell Disease Compared with History of Hb Genotype Test

Already Tested	PERCEIVED SERIOUSNESS (%) Not Serious	Uncertain	Very Serious	Total
YES	12 (37.5)	5 (22.7)	97 (40.9)	114
NO	20 (62.5)	17 (77.3)	140 (59.1)	177
Total	32	22	237	291

$\chi^2 = 2.841$, d.f. = 2, $p > 0.24$

TABLE 29

Perceived Seriousness of Sickle Cell Disease by Untested Respondents Compared with their Willingness to Undertake Hb Genotype Test

Willing to be Tested	PERCEIVED SERIOUSNESS (%)			
	Not Serious	Uncertain	Very Serious	Total
Likely	2 (10.0)	5 (29.4)	110 (78.6)	117
Uncertain/ Unlikely	18 (90.0)	12 (70.6)	30 (21.4)	60
Total	20	17	140	177

X^2 = 48.021, d.f. = 2, p < 0.00001

With respect to religion, a larger proportion (92.5%) of Orthodox Christians considered the disease very serious. This was followed by the Islamic believers (88.9%) and indigenous worshippers (77.8%). Only 50.7% of the Evangelical/syncretic Christians considered the disease serious. These differences were statistically significant as evident in Table 30.

Considering course studied, 100 percent of the health scientists believed the disease is a very serious problem, while 87.8% of biological and agricultural scientists, 82.3% of physical scientists and Engineers and 75.6% of the social science and art graduates considered it serious. Theses differences were also statistically significant as seen in Table 31.

Table 32 also shows evidence of a link between familiarity with sickle cell patients and perceived seriousness of the disease. Whereas 100 percent of respondents who has brother or sister as a patient considered the problem a very serious one. 84.7% of those who had friends as a patient believed it was serious. This was followed by 82.5% of those who had met a sickler though not related and 81.4% for those who had never met one. Of interest however, was the group who had extended family member as sickler where only 50% considered the disease as a serious problem. No link was however found between marital status and sex with perceived seriousness of the disease as can be seen in Tables 33 and 34.

TABLE 30

Religion Compared with Perceived Seriousness

Perceived Seriousness	RELIGION (%) Orthodox Christian	Islam	Indigenous African	Evangelical Christian	Total
Not Serious	12 (7.5)	6 (10.1)	2 (22.2)	34 (49.2)	54
Very Serious	147 (92.5)	48 (88.9)	7 (77.8)	35 (50.7)	237
TOTAL	159	54	9	69	291

χ^2 = 57.895, d.f. = 3, p < 0.0001

TABLE 31

Course Studied Compared with Perceived Seriousness

Perceived Seriousness	COURSE STUDIED (%) Health Sciences	Biological & Agric. Sciences	Physical Sci. and Engineer.	Social Sciences & Arts	Total
Not Serious	0 (0)	7 (12)	15 (18)	32 (24)	54
Very Serious	18 (100)	50 (88)	70 (82)	99 (76)	237
TOTAL	18	57	85	131	291

X^2 = 8.621. d.f. = 3, p < 0.035

TABLE 32

Perceived Seriousness Compared with Familiarity with Persons Having Sickle Cell Disease

Perceived Serious.	LEVEL OF FAMILIARITY (%) Brother/ Sister	Extended Family	Friend	Acquainted	None	Total
Not Serious	0 (0)	15 (50)	9 (15)	14 (18)	15 (19)	54
Very Serious	41 (100)	15 (50)	50 (85)	65 (82)	67 (81)	237
TOTAL	41	30	59	79	82	291

χ^2 = 29.859, d.f. = 4, p < 0.00001

TABLE 33

Comparison Between Perceived Seriousness and Marital Status

Perceived Seriousness	MARITAL STATUS (%) Single	Engaged/Married	Total
Not Serious	23 (10.0)	9 (15.0)	32
Uncertain	16 (6.9)	6 (10.0)	22
Very Serious	192 (83.1)	45 (75.0)	237
TOTAL	321	60	291

$X^2 = 2.082$, d.f. = 2, $p > 0.35$

TABLE 34

Perceived Seriousness Compared with Sex of Respondent

Perceived Seriousness	SEX Male (%)	Female (%)	Total
Not Serious	20 (11.0)	12 (11.0)	32
Uncertain	14 (7.7)	8 (7.3)	22
Very Serious	148 (81.3)	89 (81.7)	237
TOTAL	182	109	291

X^2 = 0.012, d.f. = 2, p > 0.99

With respect to perceived susceptibility of own children, a comparison was made between the perception of respondents who had Hb genotype test and those who have not. Analysis showed that the largest proportion (36.1%) of those believing that their children are likely to be susceptible, were found among those already tested and were found to have the abnormal haemoglobin. This was followed by those yet to undertake the test amongst whom 32.2% agreed that their children are likely. As expected, a majority (89.6%) of those already tested and had normal haemoglobin believed that their children were unlikely to be susceptible to the disease. These differences are statistically significant (Table 35).

Course studied also had positive association with perceived susceptibility of offspring. Table 36, revealed that whereas 70% of the health scientists yet to undertake the test believed their children are likely susceptible, only 43.7% of biological and agricultural scientists and 28.3% of physical scientists and engineers who are yet to be tested agreed same. For those in the Social Sciences and Arts only 25.3% of those yet to be tested believed their children are likely susceptible. Theses differences are of statistical significance as seen in the table.

There is also clear linkage between religious affiliation and the perceived susceptibility of own offspring by the respondents. Table 37 shows that the largest group believing their children are

TABLE 35

Perceived Susceptibility of Offspring with History of Hb Genotype Test

Perceived Susceptibility	HISTORY OF TESTING (%)			
	Normal Hb Gene.	Abnormal Hb Gene.	Untested	Total
Likely	7 (10.4)	17 (26.1)	57 (32.2)	81
Uncertain/ Unlikely	60 (89.6)	30 (63.9)	120 (67.8)	210
TOTAL	67	47	177	291

χ^2 = 13.390, d.f. = 2, p < 0.0013

TABLE 36

Perceived Susceptibility of Offspring by Untested Compared with Course Studied

Perceived Susceptibility	COURSE STUDIED (%) Health Sciences	Biological & Agric. Sciences	Physical Sci. and Engineer.	Social Sciences & Arts	Total
Likely	7 (70.0)	14 (43.7)	17 (28.3)	19 (25.3)	57
Uncertain/ Unlikely	3 (30.0)	18 (56.3)	43 (71.7)	56 (74.7)	120
TOTAL	10	32	60	75	177

χ^2 = 8.723, d.f. = 3, p < 0.035

TABLE 37

Perceived Susceptibility of Offspring by Untested Respondents Compared with Religion

Perceived Suscept.	RELIGION (%) Orthodox Christian	Islam	Indigenous African	Evangelical Christian	Total
Likely	36 (41.3)	11 (31.4)	2 (22.2)	8 (17.4)	57
Uncertain/ Unlikely	52 (58.7)	24 (68.6)	7 (77.8)	38 (82.6)	120
TOTAL	87	35	9	46	177

χ^2 = 8.798, d.f. = 3, $p < 0.04$

likely susceptible, are the Orthodox Christians (41.3%). This is followed by the Muslims (31.4%) and the indigenous worshippers (22.2%). Only a few (17.4%) of the Evangelical/Syncretic Christians believed their children are likely. This percentage differences are statistically significant as can be seen in the table.

Relationships are also evident in Table 38 where marital status played a major role in the perceived susceptibility of own children by the untested respondents. While a majority (74%) of those either engaged or married agreed that their children are likely to be susceptible, only a few (15.7%) of those who are still single accepted that their children are likely to be susceptible. These differences are statistically significant. Interestingly a statistical relationship was not found when familiarity with sickle cell patients was compared with the respondents' perceived susceptibility of own future offspring to the disease (Table 39).

When willingness to undertake Hb genotype test was compared with perceived susceptibility of own children, among the 177 untested respondents, analysis showed that whereas 38.5% of those who were likely to undertake the test felt their offspring were susceptible to the disease, only 20% of those who were either uncertain about taking the test or unlikely to take the test felt their offspring were likely to be suaceptible to the disease. These differences are statistically significant as seen in Table 40.

TABLE 38

Perceived Susceptibility of Offspring by Untested Respondents Compared with Marital Status

Perceived Susceptibility	MARITAL STATUS (%) Single	Engaged/Married	Total
Likely	20 (15.7)	37 (74.0)	57
Uncertain/ Unlikely	107 (84.3)	13 (26.0)	120
TOTAL	127	50	177

X^2_{Yates} = 52.122, d.f. = 1, p < 0.00001

TABLE 39

Perceived Susceptibility of Offspring by Untested Respondents Compared with Familiarity with Persons Having Sickle Cell Disease

Perceived Suscept.	LEVEL OF FAMILIARITY (%) Brother/ Sister	Extended Family	Friend	Acquainted	None	Total
Likely	3 (27)	5 (30)	13 (33)	18 (33)	18 (33)	57
Uncertain/ Unlikely	8 (73)	12 (70)	27 (67)	37 (67)	36 (67)	120
TOTAL	11	17	40	55	54	177

$\chi^2 = 0.223$, d.f. = 4, $p > 0.99$

TABLE 40

Comparison Between Perceived Susceptibility of Own Offspring and Willingness of Untested Respondents to Undertake Hb Genotype Test

Perceived Susceptibility	WILLINGNESS (%) Likely	Uncertain/ Unlikely	Total
Likely	45 (38.5)	12 (20.0)	57
Uncertain/ Unlikely	75 (61.5)	48 (80.0)	120
TOTAL	117	60	177

$\chi^2_{Yates} = 5.375$, d.f. = 1. $p < 0.03$

From the above analysis of the various hypotheses proposed by the researcher, five independent variables appear to have played major role in the outcome of the results. These include course studied, with background in health sciences being the most distinct factor positively associated with knowledge about the disease, perceptions of disease threat and willingness to be screened. Among the different religious affiliations, Orthodox Christians showed a similar positive association with the dependent variables. The presence of an sibling with sickle cell disease in the family was also found to have a strong positive link with knowledge, perceptions of disease threat and willingness to be tested.

In the next chapter, the researcher discussed the possible reasons for and implications of the influence of these variables. The value of these diagnostic findings for health education planning is also considered.

CHAPTER FIVE

DISCUSSION

This study focused on identifying the level of knowledge and perceptions about sickle cell disease and pre-marital haemoglobin genotype screening among a sample of young Nigerian graduates serving in the National Youth Service Corps. The level of awareness about the disease recorded in this study is considered high since 84.5% of the 291 respondents said they have heard about the disease. This is probably due to the high level of educational attainment of the study population. This corroborates Boroffice's (1981) findings which showed that there was correlation between educational attainment and level of awareness about this disease.

Secondary school class stood out as the single largest source of information for the respondents since as many as 75% of the sample heard at this point. The fact that 92% of those who have heard learnt about it before reaching University or Polytechnic shows that NYSC members can influence young people in the community either as teachers, health or extension workers.

However, there is discrepancy between the high level of awareness about the disease recorded among the group and the low

level of actual knowledge of the disease exhibited. The overall mean knowledge score of 5.2 (out of a possible 17 points) showed that more are aware than are properly knowledgeable about the condition. This may be due to the fact that majority of them were taught about the disease at secondary school - a point where "rote learning" rather than "critical thinking" is the vogue.

Another avenue for learning proper information about the disease, hospitals and clinics, did not rank highly as a source of information, since only 17% of those who are aware of the disease received the information from health workers. This finding shows that the situation has not changed for better since 1971 when York and Brierre (1971) reported that over-emphasis on the treatment rather than prevention of sickle cell disease by health workers makes the hospital fail in its public enlightenment role.

One major variable that enhanced knowledge was the course studied. As expected, the more related the course is to health, the more knowledgeable the individual is about sickle cell disease. This was probably why the health scientists had the highest mean knowledge score (12.7) while those in the Social Sciences and Arts, who incidentally form the bulk of the NYSC members, scored the lowest (2.5). The high score by those with health backgrounds contradicts Cohen's (1979) findings which decried the small percentage of practicing health professionals,

including senior medical and nursing students, with adequate knowledge of commonly encountered genetic conditions.

The higher scores among those with health training would incline the researcher to recommend that these NYSC members in particular be called upon to participate in sickle cell prevention campaigns. Yet, as only 6.2% of the respondents studied health sciences, reliance on them alone would be inexpedient. This is perhaps why Abramson (1973) advocated for an enlightenment programme directed towards society in general and the inclusion of persons from all walks of life in the campaign team.

The influence of religion on knowledge about sickle cell disease is most evident among the orthodox Christians, who had the highest mean knowledge score (6.5), with the Muslims following closely at 6.2 points. This is probably due to the ancient association of both orthodox Christianity and Islam with medical science. The medical missions that formed the bulk of the early and substantial proportion of the present health services in Nigeria, are provided by the orthodox Christian groups.

Evangelical and Syncretic church doctrine puts more emphasis on faith and prayers as protective and healing measures rather than on modern medical services. This has a negative implication for the readiness of members of this group to accommodate new ideas. This finding further supports Leigh's (1987) report on the

adverse effect of the teachings of certain syncretic churches which forbid members from undertaking, either in part or whole, certain medical procedures such as blood related treatments including transfusion.

The slightly better knowledge score (5.7) obtained among the males compared to 4.6 for females is probably due to the fact that society expects the male to initiate and confirm marriage proposals, and has thus saddled him with further responsibility of acquiring more knowledge on issues that will assist him in making a proper choice of spouse. This opinion is in line with earlier findings by Marini (1976) and Oppong (1981) who reported that the more topical and serious the issues affecting marriage stability are, the less knowledgeable females are about such issues. The report listed as examples of such issues genetic diseases, history of mental imbalance, sterility and barrenness. Another explanation for the score differential may be the greater access that males have to health and other science courses compared to female students.

Another important finding of this research is the higher knowledge score (6.2) obtained among unmarried respondents in contrast to 1.9 for those already engaged or introduced to their spouse and 1.7 for those already married. This gives the impression that the respondents who are yet to decide about

marriage are more responsive to information about sickle cell disease than those who have already invested in an engagement or actual marriage. This trend fulfills one of Marini's (1976) major recommendations for marriage stability. In her report on "Dimensions of Marriage Happiness," she recommended among other things, greater enthusiasm and knowledge by the unmarried, about issues affecting happiness in family life as a check against future regrets resulting from the wrong choice of spouse.

One other discovery of interest is the higher knowledge score (8.6) observed among respondents who have undertaken the test for the disease as against 3.1 recorded among those yet to undertake the test. Although it is difficult to state which aspect influenced the other, the findings could be taken to suggest that people need more knowledge about the disease in order to undertake screening.

As predicted by the Health Belief Model, there is an association between knowledge and the various measures of disease threat including seriousness and susceptibility (Rosenstock, 1974). Again, due to the cross-sectional nature of this study, it is not possible to ascertain a causal influence.

The range in knowledge score when compared with relationship of respondents to persons having sickle cell disease, showed a decreasing logical sequence. The less closely related the

respondent was to a patient, the less knowledgeable he was about the disease. A possible explanation for this trend is the fact that the more closely related one is to a patient, the more involved he is with the inconveniences resulting from the illness. This result supports earlier findings by Ojuyenwa (1980) which reported higher knowledge of handicapping conditions among mothers of deaf and blind children and lesser knowledge among neighbours of such children including the other wives of the handicapped child's father.

For this study however, whether this association between relationship to a sickle cell patient and knowledge score for the disease could be of benefit in sickle cell education will require further investigation since the percentage of sicklers in society is low. In the meantime, a major benefit derived from this association could be public enlightenment in which sickle cell patients and their close relatives are involved in education of targeted audiences about the condition. Such audiences could include school classes and social club members, so that people put a human face on the problem. Such opportunity also provides a better understanding of the disease and a firmer basis for decisions by the audience on marriage and child birth.

Furthermore, among those not yet tested, the higher knowledge score (4.0) observed among those who were likely to

undertake the screening test compared to 1.4 for those who were uncertain and 1.5 for others who said they were unlikely, suggests that higher knowledge about the disease enhances willingness to participate in the screening exercise. Earlier, it was observed, that knowledge was higher among those (114) already tested and lower among the 177 yet to undertake the test. It seems therefore, that education of the public is a major priority if screening programmes are to succeed. These results are similar to earlier findings by Fitches and Heath (1986) which recorded greater willingness to undertake prenatal diagnosis among couples who received genetic counselling and less willingness among others who were not counselled.

Of major interest, is that finding that as many as 66.6 percent of the 114 respondents already tested for the disease did so in fulfillment of higher school admission requirements. This discovery is particularly beneficial when viewed against the hope that it is possible to have all Nigerian children screened at school age if haemoglobin genotype is statutorily incorporated into school admission requirements. On the other hand, very few (4%) took the test with the intention of preventing the birth of sickle cell children. This indicates that people may engage in a recommended preventive action but not necessarily for the intended purpose. Thus in this case they may not follow through with the

second recommended step, that is using the test results to guide future marriage and child bearing plans.

Again at another level of action it was noted that awareness is not equal to knowledge nor is knowledge equal to compliance with a recommended preventive measure. Whereas 84.5 percent of the respondents said they have heard about the disease, their knowledge about the condition was quite low with a mean score well below half of the total potential points. Also, only 39.2 percent actually had ever participated in screening. Even the most knowledgeable (the health science graduates) had not participated much better (44.4% already screened) in screening than the less knowledgeable Social Science and Arts graduates (42.7%).

The general, Western religion seems to have positively influenced screening records of the respondents with orthodox Christians taking the lead. An earlier explanation may be applicable here also for this trend. They are probably more in tune with Western medicine which was also brought by the missionaries to Nigeria.

Ironically, it is those respondents who are already married or are either engaged or introduced to their future partners that have the lowest records (18.2% and 16.3% respectively) of having been screened. One would have hoped that these respondents would have had at least the same screening record rates as obtained

among the single respondents since they may have already made commitments about child bearing.

Furthermore, there seems to be an increasing motivation to undertake screening depending on how closely related one is to a person having sickle cell disease. The closer the relationship, the more likely it is that the individual would have taken the screening test. While 73.2 percent of those whose brother or sister have the disease have undertaken the test, only 31.6 percent of those who are merely acquainted with sicklers have been tested. Whether this finding is coincidental or a true evidence of the natural motivation to verify one's status regarding the disease, would require further investigation particularly as it was noted earlier that two-thirds of those already tested claimed they undertook the test in fulfillment of higher school admission requirements with only 4.3 percent claiming to have done the test out of interest. In a similar vein Table 31 showed that there was no significant relationship between perceived seriousness of the condition and history of haven been screened.

However, the influence of perception on willingness to take appropriate preventive health action is evident in Table 32 where 78.6% of respondents who perceived the disease as "very serious" said they are likely to undertake the screening test, whereas only 10% of those who claim the disease was not serious said they were

likely to undertake it. To the Health Belief Model (Rosenstock, 1974), people's likelihood to undertake preventive health action depends largely on how they perceive the disease condition. This finding is similar to earlier report by Manley (1977), which identified perceived seriousness as the major determinaot of mothers' reporting of deafness in pre-school children. In the report, 92% of 23 mothers who perceived deafness in their children as a "very serious" from the onset sought medical advice within seven days, whereas only nine percent of 16 mothers who perceived the condition as "mild" sought medical attention within the same period.

The role of course studied as a modifying factor to the willingness of the untested respondents to undertake the screening test is worthy of mention. That willingness to undertake test is higher (100%) among the health science graduates, suggests that better understanding of the condition influences desire for the screening test positively. This finding corroborates Pack (1983) who reports that better understanding of sickle cell disease maximized compliance with pre-natal diagnosis among 38 at risk, Jamaican couples. Since not all students can study the health sciences, it is necessary to develop general level courses on personal health for all University and Polytechnic students that include issues like sickle cell disease.

Willingness to undertake the test is one major area where sex makes a difference. That 73.4% of males as against 53.1% of females claimed they were likely to undertake the test suggests a situation in which the females' anxiety about securing husbands might make them reluctant to learn their screening results. One would have thought that since child bearing and care rest more heavily on them, women would want to avoid the burden of being saddled with a sickle cell child.

Religion again showed that the more cosmopolitan groups, orthodox Christians and Moslems, are more receptive to the idea of undertaking screening for the disease than others. While 85.1 percent of orthodox Christians and 71.4% of Moslems claimed they were likely to undertake the screening test, only 32.6% of Evangelical/Syncretic Christians and 33.3% of indigenous worshipers said they were likely. Again, Leigh's (1987) findings on the adverse effect of the teachings of certain Evangelical/ Syncretic churches that forbid members from undertaking certain blood related medical procedures may also explain this trend. It is no wonder therefore that religion continues to remain a major modifier of the respondents' perceived seriousness of the condition as well as of their willingness to undertake the screening test.

Also, that 91.3% of single respondents as against only two

percent of those engaged or married claimed they are likely to undertake the screening test gives the impression that people who have already invested in a relationship are less likely to succumb to procedures, the results of which could negatively affect marriage happiness. This result supports Arasomwan's (1987) findings which indicated that marriage in a typical African society is predicated much more on social consensus rather than on external criteria such as medical tests. This finding further underscores the need for the mobilization of the traditional African communities if the campaign against sickle cell disease is to succeed.

Furthermore, the personal relationship with a person having sickle cell disease continues to show its influence on willingness to undertake the screening test. While 100 percent of those having brother or sister who is patient were likely to seek screening, only 47.3% of respondents who were merely acquainted with sicklers were likely. A probable reason for this trend is the fact that personal involvement of the respondent in the care of the patient could create fear which could instigate him to want to rule out the possibility of himself being at risk of having offspring with such a dreaded disease. One possible benefit of this finding is that this group of persons, i.e. those strongly related to sicklers, could form the core of sickle cell

educational campaign teams because of their experiences through personal encounter with sickle cell individuals and their belief in the benefit of screening.

As expected also, those respondents already tested for the disease and found to be trait carriers are a bit more suspicious of the potential spread of the disease to their own offspring. Whether this suspicion will positively influence their readiness to abide by recommended preventive measures will require further investigation. However, it is gratifying to note that 78.9% of the untested respondents who said their offspring were "likely" to be susceptible to the disease also agreed that they were "likely" to seek screening.

CONCLUSIONS

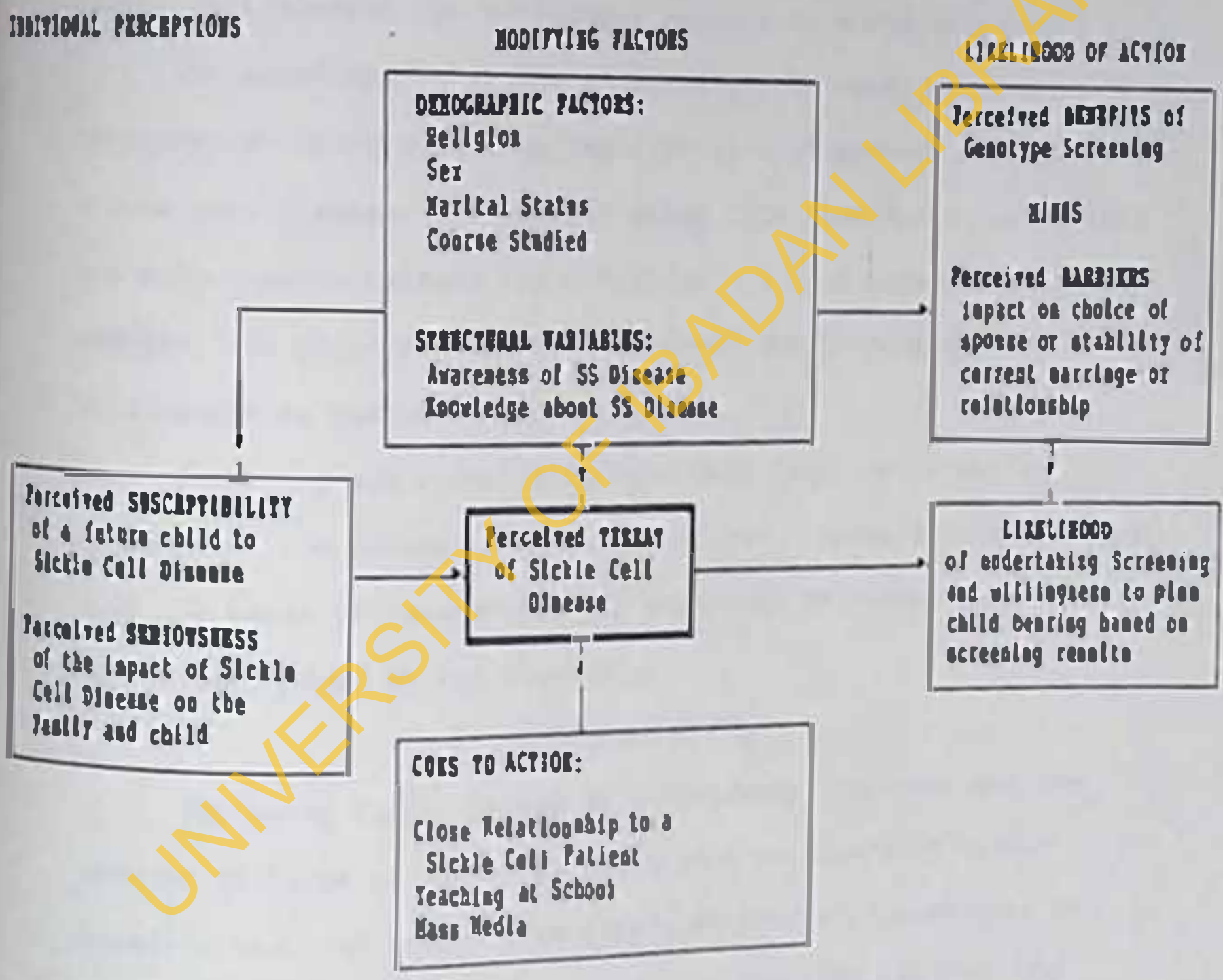
An attempt has been made in this study to find out if NYSC members' knowledge of sickle cell disease and aspects of prevention such as pre-marital haemoglobin genotype screening is adequate. It has been found that although majority of the young graduates are aware of the disease, very few (6.2%) are properly knowledgeable about the condition. As the respondents in the study are all graduates, it is likely that general awareness and knowledge about the disease among this age group is likely to be much lower than observed here.

Various reasons have been offered to explain the differences

observed according to the demographic characteristics of the study group. The positive influence of cosmopolitan religion on knowledge about the disease and willingness to be screened was noted, with the interpretation that these groups may also be more in tune with modern medical science. The low level of knowledge and reluctance to participate in screening among those who are married or engaged was explained by their commitment to a relationship, the stability of which may be threatened by screening results. The higher knowledge of male respondents was viewed in light of their socially expected role of taking the lead in making major decisions about marriage.

The major findings of this study as shown in Figure 11 which is an adaptation of the Health Belief Model (Rosenstock, 1974), and suggests a situation where an influential segment of the not too large literate population of the Nigerian Society are aware but not knowledgeable about an incurable but preventable disease. Also, women who in the society bare greater responsibility for the production of a healthy child, appear to be reluctant to know their haemoglobin genotype, possibly to avoid rejection by the male counterpart.

In addition, the results showed that the more related the academic discipline is to health, the higher the knowledge and greater the willingness of the individual to seek screening for



(Adapted from ROSENSTOCK. 1974)

Figure 11: APPLICATION OF THE HEALTH BELIEF MODEL TO THIS STUDY

the disease. The observed correlation between knowledge and perception as well as between knowledge and willingness to seek screening is similar to Lane and Scott's (1969) findings among Negroes of Richmond (U.S.A.) where the level of knowledge of sickle cell disease was strikingly related to academic course.

The major cue to action observed in the study was the respondents' relationship or familiarity with persons having sickle cell disease. As earlier noted, the findings revealed that the more closely related the individual is to a person having the disease, the greater his knowledge about the disease and his willingness to undertake screening.

Screening and counselling have been found to be useful weapons for the prevention of this disease. These and other ideas form the basis of recommendations suggested as means of solving the problem posed by the condition.

RECOMMENDATIONS

The heavy burden placed on individuals, families and the society at large by the sickle cell problem, compared to the benefits that can accrue from increased levels of awareness in general and of knowledge in particular, form the basis of the following recommendations:

1. With the school entry point having been identified as an opportunity where all children can be effectively screened

for the disease, the researcher suggests that presentation of haemoglobin genotype screening result should become a major criteria for admission into all levels of educational institutions and for recruitment into the military and para-military professions.

2. Similarly, the proposed National Identity Card, and indeed all identify cards and drivers' licenses, should contain a column for haemoglobin genotype result to encourage participation in screening.

3. Since it has been found that people do not participate in such mandatory programmes to acquire and use knowledge about their genotype, it is strongly recommended that education and counselling be incorporated into any screening activity.

4. Specific lectures in genetic diseases with particular emphasis on sickle cell disease should be included in both secondary and post secondary educational curriculum to raise the level of awareness and knowledge of the youth about the condition.

5. Sickle cell disease should be included in lectures on personal and community health at NYSC orientation camps. For a multiplier and lasting effect, all NYSC members vacating the orientation camp for their primary assignment in the communities should be given a consignment of leaflets

on sickle cell disease for distribution.

6. The apparently low level of information on sickle cell disease emanating from health workers should be rectified. It is therefore recommended that a general re-orientation of existing health workers be carried out to remind them of their responsibility in the education of the public about the condition.

7. Since women showed less likelihood to undertake the screening test, greater attention should be directed to gender specific female clubs and associations by sickle cell public enlightenment teams to ease dissemination of information and to create lasting knowledge and perception of the disease among the womenfolk.

8. In view of the low level of knowledge and the high degree of unwillingness to seek screening observed among the Evangelical/Syncretic Christians, there is an urgent need to involve more religious leaders, particularly of Evangelical/ Syncretic groups, in the enlightenment programme against the disease.

9. Finally, from health education planning perspective, involvement of sickle cell patients and their relatives in public enlightenment will assure that people put a human face into the problem.

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APPENDIX 1

QUESTIONNAIRE

This questionnaire is designed to assess Youth Corps members knowledge, and perception about sickle cell disease and Pre-marital haemoglobin genotype screening. It will be appreciated if you will provide honest answers to the questions. Results of the study will be used for research purposes only. Confidentiality of responses is guaranteed since names are not required.

Please fill in the space provided or tick box as appropriate for both Section A and B.

SECTION A

1. Age in Years: _____

2. Sex:

   Male

   Female

3. Religion: (Specify denomination)

   Orthodox (_____)

   Evangelical/Syncretic (_____)

   Muslim

   Traditional

   Others (specify) _____

4. Ethnic Group_____ State of Origin_____

5. Type of Higher Institution Attended:

   University

   Polytechnic

   School of Agriculture

   Others (specify) _____

6. Course Studied and Degree/Certificate obtained_____

---

7. Marital Status:

Single

Engaged/Introduction

Married

SECTION B

8. Have you ever heard of Sickle cell disease?

Yes

No

IF NO, SKIP TO QUESTION 13

9. When did you first heard of it?

At Primary School

At Secondary School

After Secondary School but before Post Secondary School

At the University or Polytechnic

10. Where or from whom did you hear of it first?

---

11. Since that time from where else have you hard about it?

---

12. Tick all sources of information from where you have heard of sickle cell disease.

Friends

Health Worker

Newspaper/Magazines

Class at School

Special Seminars

Relatives

TV/Radio

Others (specify) _____

13. What is sickle cell disease?

_____

_____

_____

14. What is sickle cell trait?

_____

_____

15. How is sickle cell trait and sickle cell disease transmitted?

_____

_____

_____

16. List the symptoms of sickle cell disease known to you

_____ _____ _____

_____ _____ _____

_____ _____ _____

17. Is there prevention against this disease?

Yes

No

If Yes, what is the prevention?

_____

18. Is there any impact on family when a child with sickle cell disease is born?

Yes

No

If Yes, what kind of impact?

_____

_____

19. Is there a cure?

Yes

No

If Yes, what is the cure?

_____

_____

_____

20. How would you go to confirm whether you have sickle cell disease or trait?

_____

_____

21. At what stage in ones life is it best to be tested for sickle cell trait?

_____

_____

22. What is the benefit of knowing whether you have sickle cell disease or trait?

_____

_____

23. Do you already know your hemoglobin (blood) genotype?

Yes

NO

IF NO, SKIP TO QUESTION 26

24. Which of this type do you belong

AA

AS

AC

SC

SS

Betal S-thal

Others (specify) _____

25. What prompted you to undertake the test to find out your genotype?

_____

_____

_____

IF YOUR ANSWER TO QUESTION 23 IS NO, ANSWER QUESTION 26

26. Do you intend to undertake the test to find out your genotype?

I definitely intend to take the test

I am not sure whether I will take the test

I definitely do not intend to take the test

Please give reasons for your answer above _____

_____

27. How serious is sickle cell disease?

Not at all serious

Not certain

Why? _____

_____

_____

52. If you were a carrier of the sickle cell trait (but not a sickler yourself)

a. Would you marry a spouse who refused to have his/her blood genotype screened?

Yes

No

Un decided

Give reason for your answer

_____

_____

_____

b. Would you marry a spouse if he/she was screened and also found to carry the trait?

Yes

No

Un decided

Give reasons for your answer

_____

_____

_____

53. Would you advise that Government should legislate that premarital hemoglobin (blood) Genotype screening be made compulsory to all un-married members of Society?

Yes

No

Give reasons for your answer

_____

_____

_____

34: What would you consider the major obstacles preventing people from undertaking hemoglobin (blood) genotype test?

List in other of priority

1. _____

2. _____

3. _____

4. _____

5. _____

6. _____

APPENDIX 2

DEFINITION OF TERMS

ANAEMIA:- Lack of enough blood or poor condition of the blood causing paleness.

ANOXIA:- Insufficient oxygen available for normal respiratory metabolism.

ALLELES:- One of two or more contrasting genes situated at the same locus in homologous chromosomes that determine alternative characteristics in inheritance.

PERSONNEL:- All corps members who studied health related courses.

CRISIS:- This is generally the deciding point of a disease, from which the patient either begins to recover or sinks rapidly. In sickle cell disorder, it is characterised by increased severity of the characteristic symptoms such as anaemia and pains in the bones and joints.

DOMINANT GENE:- A gene is dominant when it is expressed when heterozygous.

ERYTHROCYTES:- Red blood cells.

GENOTYPE:- The genetic make up of an individual with regard to a given pair of alleles such as AA, AS, SS, AC and SC. The alleles are the variant of a gene - A,S,C are alleles of the haemoglobin gene.

HAEMOGLOBIN:- The respiratory pigment in red blood cells.

HAEMOGLOBINOPATHIES:- A haematologic disorder caused by alteration in the genetically determined molecular structure of haemoglobin. It results in a characteristic complex of clinical and laboratory abnormalities and often sometimes, anaemia.

HAEMOGLOBIN ELECTROPHORESIS:- The most important test for sickle cell disease. It tells exactly what kind of haemoglobin the individual have. Similar results may be obtained from this test in respect of haemoglobins S and D hence there may be need to change the types or chemical used in order to distinguish one from the other. Also, the test may be used together with sickle and solubility test to help distinguish the results.

HETEROZYGOUS:- Having different alleles at the same locus. For example, an ZHb AS individual is heterozygous for normal haemoglobin (Hb A) and sickle haemoglobin (Hb S). Heterozygote is the noun form.

HOMOZYGOUS:- Having identical alleles at one locus. For example, an Hbss individual is homozygous for sickle cell haemoglobin (Hbs). Homozygote is the noun form.

MUTATION:- This is a sudden heritable change in the molecular structure of the gene due to environmental factor called mutagens. Mutation alters the properties of a gene and of its traits. The normal haemoglobin gene is the A form but mutation causes the produc-

tion from the A alleles such as S,C,D, hence they are called mutant haemoglobin variants.

NEGRO:- An individual of black African ancestry.

NUCLEOTIDE:- A monomer unpolymerized consisting of a base, a pentose sugar and a phosphate group. A nucleic acid molecule consists of many such nucleotides held together by sugar phosphate bonds.

PHENOTYPE:- This is the entire physical, biochemical and physiological nature of an individual as determined by his genotype after due interaction with his environment.

POLYPEPTIDE CHAIN:- A chain of amino acid held together by peptide bonds between the amino groups of one and the carboxyl group of an adjoining one. A protein molecule may be composed of a single polypeptide chain or of two or more identical or difficult polypeptides.

RECESSIVE GENE:- A trait is recessive if it is expressed only in individuals homozygous for the gene concerned.

SICKLE CELL ANAEMIA:- A kind of sickle cell disease which occurs when a sickle cell gene is inherited from each parent. Almost all of the haemoglobin in the blood is Hbs: no normal adult haemoglobin (HbA) is produced.

SICKLE CELL DISEASE:- This include all hereditary and haematological disorders in which sickle cell haemoglobin (Hbs) is present. It involves the possession of

two abnormal allelomorphic genes related to haemoglobin formation, at least one of which is the sickle cell gene. The genotype constituting sickle cell disease includes SS,SC,S - Thal, SE,SF - hign gene and SO (Diggs 1965, Konotey Ahulu 1974).

SICKLE CELL THALT:- This occur when a sickle cell gene is inherited from one parent and a normal gene from the other; this condition is usually without clinical significance. About 30 - 45 percent of the individuals haemoglobin is Hbs, the reminder being largely Hb A.

SICKLING TEST:- It is used to determine the existence of sickle haemoglobin in ones blood. It is carried out by mixing a drop of blood with a reducing agent on a glass slide covered by a smaller piece of glass and sealed such that oxygen does not reach the red blood cells in the drop. It does not distinguish between sickle cell disease and sickle cell trait.

SOLUBILITY TEST:- This test is based on the principle that haemoglobin S is less soluble than haemoglobin A when mixed with special chemicals in small glass tubes. It has similar drawbacks as the sickling test.

APPENDIX 3

SCORING SYSTEM FOR KNOWLEDGE ABOUT SICKLE CELL DISEASE

SERIAL NUMBER OF QUESTION	ASPECT OF KNOWLEDGE SOUGHT	MAXIMUM SCORE
13	Definition of sickle cell Disease (SCD)	2
14	Definition of sickle cell trait (SCT)	2
15	Mode of transmission of sickle cell and trait	1
16	Symptoms of sickle cell Disease	6
17	Mode of prevention of sickle Disease	2
19	Whether there is a cure for sickle Cell Disease	1
20	How to confirm ones sickle cell status	1
21	The appropriate time for healthy adults to confirm their sickle	1
22	The benefit of knowing ones sickle cell status	1
	Total score	17