UNIVERSITY OF IBADAN, IBADAN, NIGERIA

POSTGRADUATE SCHOOL

http://www.postgraduateschool.ui.edu.ng

DEAN
Professor A.A. Adeyemo, Ph.D (Ibadan)
Mobile: 08023749637
E-mail: adeyemi@yahoo.com
or adeyemio@mail.ui.edu.ng

SUB DEAN (Sciences)
Dr. Oyedunlan A. Adedoyin
DVM, Ph.D (Ibadan), MCVSN
Mobile: 08023222926
E-mail: oye.adedoyin@yahoo.com

DEPUTY REGISTRAR/SECRETARY
A.A. Olade, B.A., DPA (Ife)
Mobile: 08032316456
E-mail: aa.olade@gmail.com
or olade@mail.ui.edu.ng

SUB-DEAN (Arts & Humanities)
A.I. Alarape, Ph.D (Ibadan)
Mobile: 08050884273
E-mail: a.alarape@gmail.com
or a.alarape@mail.ui.edu.ng

...centre of excellence
for postgraduate training and research

Ref. No: S.I. 168460

Date: 01 September, 2016.

Adeleke Anthony ADEOLU

Dear Mr/Mrs ADEOLU,

Registration of Title of Dissertation

I write to inform you that the Executive Committee of the Postgraduate School at its reconvened meeting held on 16 June 2016 approved the recommendation from the Faculty of Public Health for the registration of your M.P.H. Dissertation titled:

KNOWLEDGE,PERCEPTION AND ATTITUDE OF ABOUT-TO-WED COUPLES TOWARDS PREMARITAL SICKLE CELL GENETIC TESTING AND COUNSELLING IN IBADAN NORTH-EAST LOCAL GOVERNMENT REGISTRY, OYO STATE

The approval is with effect from 4 September, 2016. The registration would lapse if not examined by 16 December, 2016.

Yours sincerely,

Modupe G. Bada (Mrs.)
Committees Officer
for: Deputy Registrar/Secretary

Cc: Dean,
Faculty of Public Health
Head,
Department of Health Promotion and Education
Examinations Officer, Postgraduate School.
Records Officer, Postgraduate School.
Student Copy.
File Copy.

KNOWLEDGE, PERCEPTION AND ATTITUDE OF ABOUT –TO-WED COUPLES TOWARDS PREMARITAL SICKLE CELL GENETIC TESTING AND COUNSELLING IN IBADAN NORTH-EAST LOCAL GOVERNMENT REGISTRY, OYO STATE.

BY

ADELEKE, ADEOLU ANTHONY

MATRIC NO.: 168160

B. Tech Human Physiology (Ladoke Akintola University of Technology)

A Dissertation in the Department of Health Promotion and Education Submitted to Faculty of Public Health, College of Medicine, in Partial Fulfilment of the requirements for the award of Degree of Master of Public Health

of the

UNIVERSITY OF IBADAN.

JANUARY, 2017.

DEDICATION

This work is dedicated to Almighty God, the Alpha and Omega.

To my Parents, Mr. and Mrs. A.A Adeleke.

To my wife, Olajumoke Adeleke.

To my children, Richard Adedamola and Michelle Aderinsola Adeleke.

And most of all, families battling with the scourge of Sickle Cell Disease.

CERTIFICATION

I certify that this project was carried out by ADELEKE, Adeolu Anthony of the Department of Health Promotion and Education, Faculty of Public Health, College of Medicine, university of Ibadan, under my supervision.

Supervisor

PROFESSOR OLADIMEJI OLADEPO

B.Sc. (Hons), MPH (Ibadan), FRSPH (UK).

Department of Health Promotion and Education,

Faculty of Public Health, College of Medicine,

University of Ibadan,

Ibadan, Nigeria.

ABSTRACT

Sickle cell disease affects millions of people throughout the world and is particularly common among African descents. Earlier studies in Oyo State focused on attitude towards mandatory premarital testing with no information on knowledge, perception and attitude of about-to-wed couples towards premarital sickle cell counselling and testing. This study was therefore designed to investigate the level of knowledge, attitude and perception of about-to-wed couples towards Premarital Counselling and Testing (PSCCT) in Ibadan North-East Local Government Area (LGA) of Oyo State.

The study design was descriptive. A three-stage sampling technique was used to select LGA, registry, and 400 consenting respondents. A semi-structured, interviewer-administered questionnaire was used to obtain data on respondents' socio-demographic characteristics. Knowledge was measured on a 30-point scale on which scores of ≤ 10, 10 - 20 and > 20 were categorised as poor, fair and good, respectively Perception towards PSCCT was measured on a 20-point scale; scores of ≤10 and >10 were classified as negative and positive, respectively. Attitude was measured on a 20-point scales on which scores of ≤10 and >10 were classified as negative and positive, respectively. Quantitative data were analysed using descriptive statistics, Chi-square test and logistic regression at p=0.05. Seven In-depth Interview (IDIs) were conducted on PSCCT and was analysed thematically.

Age was 31.7±5.5 years, 55.0% were males, 98.0% had formal education, 52.0% were Christians and 62.0% were self employed. Majority (81.8%) had positive perception about PSCCT. Respondents' knowledge score was 11.4±5.0 with (63.0%) having a fair knowledge. Reported purposes of PSCCT were; making an informed marital choice (28.8%) and preventing incidence of sickle cell disorder among offspring (24.0%). Majority (91.5%)

ABSTRACT

Sickle cell disease affects millions of people throughout the world and is particularly common among African descents. Earlier studies in Oyo State focused on attitude towards mandatory premarital testing with no information on knowledge, perception and attitude of about-to-wed couples towards premarital sickle cell counselling and testing. This study was therefore designed to investigate the level of knowledge, attitude and perception of about-to-wed couples towards Premarital Counselling and Testing (PSCCT) in Ibadan North-East Local Government Area (LGA) of Oyo State.

The study design was descriptive. A three-stage sampling technique was used to select LGA, registry, and 400 consenting respondents. A semi-structured, interviewer-administered questionnaire was used to obtain data on respondents' socio-demographic characteristics. Knowledge was measured on a 30-point scale on which scores of ≤ 10, 10 - 20 and > 20 were categorised as poor, fair and good, respectively Perception towards PSCCT was measured on a 20-point scale; scores of ≤10 and >10 were classified as negative and positive, respectively. Attitude was measured on a 20-point scales on which scores of ≤10 and >10 were classified as negative and positive, respectively. Quantitative data were analysed using descriptive statistics, Chi-square test and logistic regression at p=0.05. Seven In-depth Interview (IDIs) were conducted on PSCCT and was analysed thematically.

Age was 31.7±5.5 years, 55.0% were males, 98.0% had formal education, 52.0% were Christians and 62.0% were self employed. Majority (81.8%) had positive perception about PSCCT. Respondents' knowledge score was 11.4±5.0 with (63.0%) having a fair knowledge. Reported purposes of PSCCT were; making an informed marital choice (28.8%) and preventing incidence of sickle cell disorder among offspring (24.0%). Majority (91.5%)

wanted religious institutions to encourage PSCCT among intending couples. Respondents' attitude score was 7.1±1.8. Perception score was 12.4±3.8. Many (61.5%) had used PSCCT service, most of whom used health facilities. Most, (58.0%) spent between ₦300 - ₦500 for the test and majority (91.5%) said marriage institutions should encourage their members to go for PSCCT. More respondents with formal education (98%) significantly utilised PSCCT compared to those that were not educated (2%). Respondents with formal education were more likely to utilise PSCCT compared to those that were not educated (OR=4.2; CI=1.5-10.4). (61.5%) of respondents that utilised PSCCT had good attitude towards PSCCT compared to those that did not utilise PSCCT. Utilisation of PSCCT was significantly associated with Knowledge of PSCCT. Respondents with fair knowledge were more likely to utilise the PSCCT compared to those with poor knowledge (OR: 2.5, CI=2.3-4.2). Respondents' perception score was 12.4±3.7. The IDIs revealed that majority used the hospital, School clinic and private laboratory for PSCCT.

The good knowledge, attitude and perception of respondents towards PSCCT suggest the acceptability of the services among intending couples. Therefore, promotion of premarital sickle cell counselling and testing at marriage registries and all government approved churches for weddings is therefore recommended

Keywords: Sickle cell disease, premarital, genetic counselling and testing, religious institutions.

Word count: 468

ACKNOWLEDGEMENT

This study would not have been possible without of my professors, family, mentors and friends. I must thank all of my lectures, most importantly my supervisor Prof. Oladepo. I am also indebted to the crop my outstanding lectures; Prof Ajuwon, Oyedunni O, Arulogun, Dr. Fredrick Oshiname, Dr. Oyediran Oyewole, Dr.Titiloye Musbau, Dr. Akpan, Dr. Modupe O, Oyetunde, Mrs. Adeyinka Desmenu, Mrs. Mojisola Oluwasanu, Mr. Femi Dipeolu, Mr John Imaledo. I say thank you all.

To my family, thank you for encouraging me in all of my pursuits and inspiring me to follow my dreams. I am especially grateful to my parents, Mr and Mrs A.A Adeleke who supported me emotionally and financially. I have always known that you believed in me and wanted the best for me. To my siblings; Mrs. Abosede Ige, Fr Albert Adeleke, Ademola, Adebambo, Nurse Joke and Adebowale, thank you for your supports in your own way. Thank you for teaching me that my job in life is to learn, to be happy, to know and understand myself; only then could I know and understand others.

To my wife, Mrs. Olajumoke Adeleke for your unfounded efforts during the MPH programme. I say a big thank you. To m children, Richard Adedamola Ademurewa and Michelle Aderinsola Adeleke for your understanding during the period of financial commitment.

I owe a debt of gratitude to the administrative staff; Mr Olubodun, Mr Lanre, Mr Bello, you authored my coming for MPH may the good Lord bless your children. My appreciation also goes to my special friend Toriola Yusuff for your selfless efforts during the course of this research. I will forever be grateful to you.

ADELEKE, Adeolu Anthony.

TABLE OF CONTENTS

LIST OF TABLES

LIST OF FIGURES

LIST OF APPENDICES

LIST OF ABBREVIATIONS

HIV: Human Immunodeficiency Virus

MPH: Master of Public Health

M.SC: Master of Science

OND: Ordinary National Diploma

PSCCT: Premarital Sickle Cell Counseling and Testing

RA: Research Assistant

SCD: Sickle Cell Disorder

SPSS: Statistical Package for Social Sciences

WHO: World Health organisation

PMS: Premarital Screening

LGA: Local Government Area

ATW: About-to-Wed

IDI: In-depth Interview.

OPERATIONAL DEFINITIONS

Knowledge

It is the information, understanding or skills that one acquires from experience or education. Knowledge in this study focuses on respondents' level of understanding of sickle cell disorder.

Attitude

This is the opinions and feelings towards a phenomenon. This study will focus on level of perception towards premarital sickle cell counselling and testing.

Perception

This is the view or understanding towards a phenomenon. This study will focus on level of perception towards premarital sickle cell counselling and testing.

Registry

A service provided that assists engaged couples in documenting their marriage contract.

About-to-wed

These are two consenting adults that are preparing for their wedding.

CHAPTER ONE

INTRODUCTION

1.1 Backgrouod Knowledge

Genetic testing and counselling is a very crucial aspect of human lives as having details about one's genetic makeup may help individual make an informed decision about his or her marital life. Genetic counselling is the processes by which patients or relatives who are at risk of an inherited disorder, are advised of the consequences and nature of the disorder, the probability of developing or transmitting it, and the options open to them in management and family planning in order to prevent, avoid or ameliorate it (Dipika and Kishalaya, 2011). Genetic counselling provides the link between genetic technologies, several of which have been acquired through the Human Genome Project, and patient care. It can be defined as a communication process which involves diagnosis, explanation and options. Therefore; genetic counselling is branched into diagnostic (the actual estimation of risk) and supportive aspects.

The sickle haemoglobin (HbS) gene was developed in response to severe malaria endemicity (Piel etal; 2010). It has been determined that up 24% of Nigerians have the sickle cell gene (WHO, 2006). Multiple studies have shown that the sickle cell gene is indeed protective against malaria, as heterozygotic persons with a single sickle cell gene (AS individuals) have been shown to have a decreased risk for the disease (Rees etal; 2010, Williams and Obaro, 2011). Homozygotic individuals (SS individuals) that inherited two HbS alleles from both parents are however not only at greater risk of contracting malaria, but are also at higher risk of several health problems, linked to the tendency of the altered red blood cells to become sickled, and then stick to blood vessels, causing ischaemia to multiple cells and organs of the body, and resulting in the sickle cell disease (Rees etal; 2010, Williams and Obaro, 2011). Many religious organisations are currently requesting about-to-wed couples to conduct pre-marital genotype test. It is believed that marriage between two carriers of Sickle Cell Trait (HbAS) could only be described as an irrational plunge into troubles and as such, the couple risks the tendency to produce children with sickle cell anaemia in every four children they have. Mendellan principle states that, there is the tendency that two people who are carriers could give birth to sickle cell anaemia child/children.

Of all genetic disorders, this study will focus only on sickle cell disease because it is the commonest genetic disorder in this part of the world. Thus, this study aimed to unveil the knowledge, attitude and perceptions of about to wed couples because they are the population that mostly go for premarital genetic testing and counselling and to provide useful

information for appropriate methods and strategies to promote premarital counselling and testing in Ibadan North-East Local Government Area.

1.2 Statement of the Problem

Sickle cell disease affects millions of people throughout the world and is particularly common among those whose ancestors came from Africa (CDC, 2011). The World Health Organization (2010) documents that sickle cell disease is a hereditary genetic disorder that occurs when a person has inherited two abnormal mutant haemoglobin (HB) genes from both parents. It occurs at a frequency of 1 out of 1600 among black people (Adeyemo et al; 2007). It contributes to the equivalent of 5% of under-five deaths on the African continent (WHO, 2005). Nigeria accounts for 50% of sickle cell disease (SCD) births worldwide and about 2.3% of her population suffers from SCD with 25% of Nigerians being healthy carriers (Oludare and Ogilli, 2013). Many also die before their reproductive age. Sickle cell anaemia poses serious health concern, especially in developing countries. Complications of sickle cell disease include serious infections, damage to vital organs, stroke, kidney damage, respiratory problems, bone marrow failure, growth failure, cognitive impairment, maturational delay in children as well as high maternal and foetal morbidity and mortality (Zemel etal;2007, Ocheni etal;2007). Awareness on genetic understanding and screening is not a common practice and the diagnosis is usually made when presented with a severe complication (Adeyemo et al; 2007). Even when tragedies such as two or more miscarriages, still births, or children die in infancy, many at times doctors do not order a blood test to take a closer look at genetic make-up of parents or refer them to a genetic counsellor.

The chronic nature of SCD requiring life-long medical attention, expensive supportive symptomatic therapy, its specialised care, the associated high morbidity, reduction in life expectancy of the affected, poor school attendance, the potential risk of developing drug addiction, especially to opiates, and its burden on the affected families all indicate that the condition is a major public health problem requiring attention (WHO, 2006). Prevention of the disease through carrier identification and genetic counselling remains the only realistic approach to reduce the impact of the disease and allows better use of available resources in the low-income countries where the condition is most prevalent. Programmes of population

screening and genetic counselling can have a major impact on the birth rate of children with SCD and other genetic diseases (Arrayed, 2005 and Vogel, 1982)

1.3 Justification of the Study

Despite the large number of people affected with sickle cell disease, the level of knowledge about sickle cell disease is still low (Ugwu, 2016). In Nigeria, various studies have reported poor knowledge of sickle cell disorder among students. A study conducted by Adewuyi (2000) among fresh University graduates in Ilorin, Nigeria reported poor knowledge of SCD, as only 43% of the respondents showed little understanding of the disease. Similar study conducted in Benin City, Nigeria by Bazuaye et al; (2009) reported that majority of the students (55.1%) do not know their genotype and only 18% had some correct idea about SCD. Knowledge gap has also been shown by a similar study conducted in Jos, Nigeria by Olarewaju et al; (2013) who reported that many of the students (25.5%) had wrong belief that SCD is caused by evil spirit. Study from Ghana has also indicated knowledge gap despite the high prevalence of sickle cell carrier status approaching 25% and the universal newborn screening program being introduce recently in Ghana (Ross et al; 2011).

In another study, comparism of the level of knowledge was made among University students in Texas and Enugu, Nigeria, reported that on the average, students are aware of sickle cell anaemia and its carrier state, but there is still some knowledge gap about SCD especially among students in non-medically-related faculties many of whom did not know their genotype (Ogamdi and Onwe, 2000). The SCD strategy for WHO African region seeks to increase individual and community awareness about SCD and strengthens primary prevention, reduces disease incidence, morbidity and mortality, and improves quality of life. The strategy also contributes towards the achievement of Millennium Development Goals 4 and 5. Knowledge about SCD is a way of preventing and controlling the scourge, since people will be better equipped to take informed decision concerning their marriage and the about-to-wed couples are good entry point for interventions aimed at controlling the disease.

Since the marriage registry is an institution that has access to folks from different religious backgrounds, it offers a good representation of Nigerian intending couples, there is need to assess the level of knowledge, attitude and perception about sickle cell disease because an understanding of these factors will help to fashion appropriate public health education programs to encourage positive attitude, perception and knowledge of the condition. The aim of this study is therefore to assess the level of knowledge, perception and attitude of abut-to-wed towards premarital sickle cell counselling and testing. It is hoped that the results of the study would help in packaging a more comprehensive and government-led premarital screening

programme in Nigeria. Finally, the result of this study will be useful for health care providers and policy makers regarding awareness and health education of potential couples concerning premarital sickle cell disease genetic testing and counselling. This will in turn enhance making informed decisions about marriage and procreation among them.

1.4 Research Questions

The following research questions were set for the study;

1. What is the level of knowledge of respondents regarding issues about sickle cell disorder?
2. What is the respondents' attitude towards pre-marital sickle cell genetic testing and counselling?
3. What is the respondents' perception towards pre-marital sickle cell genetic testing and counselling?
4. What is the respondents' level of utilisation of pre-marital sickle cell genetic testing and counselling services?

1.4.1 Broad Objective

The broad objective of the study was to determine the level of knowledge, perception and attitude of about-to-wed couples in Ibadan North-East about sickle cell premarital counselling and testing services and its utilisation to obtain and provide important baseline data.

1.4.2 Specific Objectives

The specific objectives that guided the study include, to;

1. Assess the level of knowledge regarding sickle cell disorder among participants in this study.
2. Determine the attitudinal dispositions of about-to-wed couples towards premarital genetic testing and counselling.
3. To determine the perception of about-to-wed couples towards premarital genetic testing and counselling.
4. Determine the prevalence of utilisation of premarital sickle cell genetic testing services among participants in this study.

1.5 Research Hypotheses

1. There will be a significant relationship between knowledge of sickle cell genetic disorder and utilisation of premarital sickle cell genetic counselling and testing services.
2. There will be a significant relationship between perception and utilization of premarital sickle cell genetic counselling and testing.
3. There will be a significant relationship between attitude and utilisation of premarital sickle cell testing and counselling.
4. There will be a significant relationship between level of education and utilisation of premarital sickle cell counselling and testing.

CHAPTER TWO

LITERATURE REVIEW

2.1 Premarital Screening Programmes for Haemoglobinopathy

The legal relationship between a husband and a wife, marriage can maintain good sexual reproductive health if it is safe or rather create ill-sexual reproductive health in couples, if it is unsafe (Ettie, 2005). This is partly due to the colossal ignorance of the public in regard to sex and health implication of marriage, and partly duo to the fact that marriage is mainly controlled by lawyers and priests or pastors- most of who take no regard for the health status of would-be couples and their offspring- than by would-be couples themselves and medical doctors (Ettie, 2005).

Preventive genetic services based on population screening are now an integral part of maternal and child health programmes in many parts of the world. Mass screening and genetic counselling have been carried out widely, but they are not as effective as antenatal screening programmes. New developments in DNA technology, ultrasound scanning and assaying maternal blood factors greatly increase the potential for improving human health (Kulicv and Modell, 1990)

The prevalence of inherited blood disorders in certain parts of the world is high, including the autosomally inherited haemoglobinopathies, thalassaemia and sickle cell disease. Premarital screening aims to identify carriers of the haemoglobin disorders, in order to assess the risk of having children with a severe form of disease. The couple can then choose whether or not to have an affected child.

Improved healthcare and management means that the numbers of thalassaemic patients and their life expectancy are increasing, and this places extra demands on healthcare systems, meaning that some countries are unable to deliver optimum treatment to all their affected patients. Effective prevention can maximize the available resources if it is instituted properly, preventing up to 95% of affected births (WHO, 2006). Healthy carriers of beta-thalassemia can be identified inexpensively and accurately by a simple blood test. Couples who undergo testing can be informed about genetic risks and given options for reducing risk, including prenatal diagnosis (WHO, 2006).

2.2 History of Premarital Screening for Thalassaemia and Sickle Cell Disease

Premarital thalassaemia screening was first carried out in 1975 by Silvestroni and colleagues (1978) in Latium, Italy, as part of a school prevention programme. Screening for sickle cell anaemia began before this, in Virginia in 1970, nationwide screening programmes also began in Canada, Cyprus, Greece and the UK during the 1970s, with proven success (Tosun etal; 2006) until this point, the genetic causes of haemoglobinopathy were understood, but little had been done to prevent them in newborns.

Angastiniotis and Modell (1998) classified countries who deliver premarital screening programmes into three categories:

- Endemic Mediterranean countries in which preventive programmes are long-established, with success rates (preventive) of 80-100% and optimum treatment via specialist clinics.
- Developed, industrialized countries in which prevalence is increasing because of migration; these countries can fund screening programmes, but find it difficult to reach immigrants with certain cultural backgrounds.
- Developing countries in which there are economic difficulties and other health priorities (e.g. infectious disease control), or other religious and cultural constraints.

In Cyprus, Greece and Italy, premarital screening for thalassemia has been normal practice for a long time because consanguinity is high. Similar preventive programmes have been introduced in Bahrain, China, India, the Islamic Republic of Iran, Indonesia, Malaysia, the Maldives, Singapore and Thailand, and recently in Saudi Arabia and United Arab Emirates. In the UK, Northern Ireland and other northwest European countries, prenatal diagnosis is available and abortion is a prevention strategy (WHO, 2006)

In China, couples who wish to marry are extensively tested, including physical examination. They are given premarital health instructions and 'counselling' – in the form of watching videotapes of the type of child they might conceive – after which appropriate measures are taken. This approach has been strongly criticized in terms of human rights, control, oppression and eugenics, even though the value of vigilant premarital screening is acknowledged (Hesketh, 2003).

In Lebanon, thalassaemia patients are managed in chronic-care centres in collaboration with the ministries of Social Affairs and Public Health. Their screening programme includes

providing information, training health professionals and developing training materials, with priority given to disseminating knowledge and increasing public awareness (Inati et al, 2006).

Tosun et al, (2006) studied the premarital haemoglobinopathy screening programmes in Mersin, Turkey, where consanguinity is 30%. If the man and the woman are both carriers, results are given confidentially and they are counselled about their options, including prenatal diagnosis.

2.3 Role of Culture and Education in the Success of Screening Programmes

'Consanguinity' refers to relationships by blood or common ancestry, in which the chances of offspring inheriting a recessive allele for a disease are increased; the closer the relationship, the greater the risk. Marriages between members of the same tribe or extended family group are favoured in some cultures, including those between first cousins. Consanguineous marriages are uncommon in Western countries. Marriage between first cousins is forbidden by the Orthodox Church and Roman Catholic Church, and may be seen as incestuous in the United States.

Personal characteristics including socioeconomic status have implications for the outcome of premarital screening programmes. Education of the couples who are to be screened is extremely important and it is essential to educate all members of the screening team (laboratory technologists, nurse practitioners, physicians, counsellors, outreach workers and social workers). According to Schmidt (1974), sufficient planning in the educational area before the first blood sample is drawn can avoid failures of the programme.' The meaning of the term 'carrier status' should be made known to members of the public long before they get married. For successful public education, governments and government organizations must cooperate, as well as community and religious leaders, school parent organizations and health personnel (Tosun et al, 2006)

A study by Eshra and colleagues (1989) based in Egypt revealed gaps in knowledge regarding premarital screening, even among educated people. These people acquired their knowledge from the mass media and medical personnel. People who responded to information about premarital screening had favourable attitudes towards premarital counselling and examination of consanguineous marriages, possibly relating to social changes, declining illiteracy, increasing economic pressures, increasing numbers of nuclear families and longer waiting times before starting a family. People with a negative attitude towards these tests were mostly unmarried males. Eshra and colleagues (1989) therefore suggested that education programmes about the benefits of premarital examination should

target unmarried males, so they can make informed choices about unmarried females and consanguineous marriages.

Religious beliefs restrict the success of screening programmes in some communities. In Southern Iran, premarital screening has been mandatory for 10 years, yet high-risk couples still get married and give birth to children homozygous for beta-thalassaemia. Often this is because of religious and traditional cultural restraints (Karimi etal; 2007) in the case of Islam, consanguineous marriages are permitted, so thalassemia persists in some parts of the community, making the programme redundant.

Some people believe that their fate is determined by God and therefore accept the risk of having a sick child. A recent report in The Jordan Times (Monaghan, 2008) showed that many Jordanians view the results of their 'unions' as *fate*. On the contrary, there are many teachings in Islamic culture that promote healthy marriage and the role of counselling (Albar, 1999).

Reports by Karimi etal; (2007), Monaghan, (2008) and AlKhaldi etal. ; (2007) were from three different Islamic countries, and all three provided evidence that religious beliefs could be obstacles to the success of premarital screening programmes, regardless of other factors such as education level. The same conclusions were reported long ago in other (non-Muslim) communities. In 1981, Angastiniotis and Hadjiminas, (1981) stated that support from the Church was the main reason for the success of screening programmes in Cyprus and Greece.

2.4 Genetic Counselling

Genetic counselling is the process by which an individual or a family obtains information about a genetic condition that may affect them, so that they can make appropriate decisions about marriage, reproduction and health management (Albar, 1999) Genetic counselling protects the autonomy of the couple, fulfilling their right to be fully informed about the disorder and all available options (WHO, 2006). Although premarital tests for haemoglobinopathies are reliable and useful, not everyone with these genes responds to counselling. Neal-Cooper and Scott[4] reported that young couples' concerns about producing a child with sickle cell disease are often offset by their strong desire to have children regardless of risk. The researchers suggested that at-risk couples should be contacted directly by counsellors and encouraged to undertake education and counselling.

Studies of patient perspectives about premarital examinations reveal a need for physicians to offer counselling for various health problems before the patient asks, so they can turn down unwanted help rather bring up sensitive issues (Schmitt, 1990). Screening programmes that

inform about the risks associated with child bearing should be performed well before child-bearing age. Unfortunately, premarital screening often comes too late for couples to change their opinions about marriage based on their haemoglobin phenotype, because by this point they are already committed to their relationship. Furthermore, it may be 'taboo' for a woman to reject marriage for these reasons, and it may affect her social life, preventing her from ever getting married.

One successful approach is 'solution-focused' premarital counselling. Murray and Murray (2004) discussed how this focuses on a couple's resources, helping them to develop a shared vision for the marriage. Background information about premarital counselling and solution-focused therapy provide a framework in which intervention strategies in those confirmed with positive status for a disease can be developed. These solution-oriented interventions include solution-oriented questions and feedback, as well as a Couple's Resource Map (CRM) which depicts the support available to the couple from various personal, relationship and contextual resources (Christene, 2008) The available choices include avoidance of marriage, reproductive options for those who proceed with the marriage following prenatal diagnosis, adoption of the affected child, donation of sperm, ova or a pre-embryo from an unaffected individual, and pre-implantation diagnosis (Monaghan, 2008). Choosing the best option depends on availability, cost, and local regulations and religious rules. For example, in Saudi Arabia adoption of children is prohibited on religious grounds and prenatal diagnosis is useless because abortion is forbidden unless the foetus is malformed. However, pre-implantation diagnosis is permitted and affordable.

Thus the success of genetic counselling depends on the approach adopted by the counsellor as well as the education and attitude of the couple. Screening programmes must be equitable, accessible and understood by the target population, but most importantly they must comply with the prevailing cultural, ethnic, economic and societal values.

2.4.1 Genetic Testing

Genetic testing usually involves having a sample of your blood or tissue taken. The sample will contain cells that contain your DNA and can be tested to find out whether you are carrying a particular mutation and are at risk of developing a particular genetic condition.

Genetic testing is only useful if it is known that a specific genetic mutation causes a condition. For example, a specific gene is known to cause cystic fibrosis (a condition where there is muscle weakness and progressive loss of movement). It is therefore possible to test a blood sample for the presence or absence of this gene. Some genetic conditions are caused by

inform about the risks associated with child bearing should be performed well before child-bearing age. Unfortunately, premarital screening often comes too late for couples to change their opinions about intiriage based on their haemoglobin phenotype, because by this point they are already committed to their relationship. Furthermore, it may be 'taboo' for a woman to reject marriage for these reasons, and it may affect her social life, preventing her from ever getting married.

One successful approach is 'solution-focused' premarital counselling. Murray and Murray (2004) discussed how this focuses on a couple's resources, helping them to develop a shared vision for the marriage. Background information about premarital counselling and solution-focused therapy provide a framework in which intervention strategies in those confirmed with positive status for a disease can be developed. These solution-oriented interventions include solution-oriented questions and feedback, as well as a Couple's Resource Map (CRM) which depicts the support available to the couple from various personal, relationship and contextual resources (Christene, 2008) The available choices include avoidance of marriage, reproductive options for those who proceed with the marriage following prenatal diagnosis, adoption of the affected child, donation of sperm, ova or a pre-embryo from an unaffected individual, and pre-implantation diagnosis (Monaghan, 2008). Choosing the best option depends on availability, cost, and local regulations and religious rules. For example, in Saudi Arabia adoption of children is prohibited on religious grounds and prenatal diagnosis is useless because abortion is forbidden unless the foetus is malformed. However, pre-implantation diagnosis is permitted and affordable.

Thus the success of genetic counselling depends on the approach adopted by the counsellor as well as the education and attitude of the couple. Screening programmes must be equitable, accessible and understood by the target population, but most importantly they must comply with the prevailing cultural, ethnic, economic and societal values.

2.4.1 Genetic Testing

Genetic testing usually involves having a sample of your blood or tissue taken. The sample will contain cells that contain your DNA and can be tested to find out whether you are carrying a particular mutation and are at risk of developing a particular genetic condition.

Genetic testing is only useful if it is known that a specific genetic mutation causes a condition. For example, a specific gene is known to cause cystic fibrosis (a condition where there is muscle weakness and progressive loss of movement). It is therefore possible to test a blood sample for the presence or absence of this gene. Some genetic conditions are caused by

particular mutations. Others can be caused by any mutation in a gene. For example is usually caused by a few particular mutations, which means it is relatively easy to test for.

A genetic disorder is an illness caused by abnormalities in the genome, especially a condition that is present from birth congenital. Most genetic disorders are quite rare and affect one person in every several thousands or millions.

Genetic disorders are heritable, and are passed down from the parents' genes. Other defects may be caused by new mutations or changes to the DNA. In such cases, the defect will only be heritable if it occurs in the germ line. The same disease, such as some forms of cancer, may be caused by an inherited genetic condition in some people, by new mutations in other people, and by non - genetic causes in still other people.

2.4.2 Uses of Genetic Testing and Counselling.

Among their many uses, pre-marital genetic screening most commonly present an opportunity for individuals to become informed about their genetic predisposition to disease, and for couples to be aware of the possible genetic characteristics of their unborn children. Hence, if one holds the view that one of the reasons for marriage is procreation, then worrying about genetic compatibility and avoiding genetic inheritance of grave consequence becomes something to strongly consider. According to El-Hazmi and Warsy (2011), pre-marital genetic screening is the screening of the prospective couples for a genetic disease, genetic predisposition to a disease, or a genotype that increases risk of having a child with a genetic disease. It gives the couples information about their predisposition to certain diseases and the odds of passing on those diseases to their unborn children. It is part of every couple's intelligent wedding plan that provides them with opportunity for prevention, management and treatment of diseases.

According to WHO (1999) pre-marital genetic screening is aimed at addressing the medical and psychosocial needs of affected individuals and their families through early and accurate clinical and laboratory diagnosis, coordination of the multidisciplinary and long-term management of patients, anticipatory guidance of predicted problems based on the natural history of the condition, genetic counselling and psychosocial support to the patient and family. Knowledge of pre-marital genetic screening allows a person to take steps to reduce his or her risk. For people at an increased risk of certain disorders, healthcare professionals may recommend more frequent screening starting at an earlier age. Healthcare providers may

also encourage regular checkups or testing for people with a medical condition that runs in their family. According to WHO (2006), methods of preventing genetic diseases include pre-marital screening and genetic counselling, prenatal diagnosis, preconception diagnosis and implantation of normal embryos after in-vitro-fertilisation, and in-uterotherapy using stem cell transplantation. Prevention of the disease through carrier identification and genetic counselling remains the only realistic approach to reduce the impact of the disease and allow better use of available resources in the low-income countries where the condition is most prevalent.

2.5 Sickle Cell Disorder

The haemoglobinopathies are autosomal recessive defects of these genes. Over 600 different varieties have been described (Huisman et al; 1993). They affect either the structure of β-Hb (the variant disorders, e.g. sickle) or reduce the quantity of either α- or β-Hb chains (the thalassaemias). Sickle Hb (S) is a qualitative defect in which a DNA substitution in the β-chain results in an alteration at position 6 of the amino acid chain, giving rise to a structurally different Hb when the S chains are assembled in the Hb molecule. This variant alters the electric charge of the molecule, thus giving rise to clinical pathology and allowing its easy detection in the laboratory.The thalassaemias are named after the chain that is deficient (i.e. α- or β-thalassaemia). The former is usually due to gene deletions, the latter to non deletional alleles, of which over 100 have been described (Higgs,1993;Thein,1993). Clinically severe conditions occur when either both β-genes, or three or four α-chains are affected.

a) Transmission

A distinction must be made between carriers (who have only one affected globin locus and remain healthy throughout life, but are at risk of transmitting the disease to their descendants) and people who are homozygous, or doubly heterozygote, for a disorder. The conditions are recessively transmitted according to Mendelian genetics. Parents have a one in four risk of conceiving an affected child if both parents are carriers.

b) Beta-thalassaemia

Beta-thalassaemia major is characterised by deficient or absent β-chain production and extra-medullary erythropoiesis. Raised levels of Hb F compensate partially but death occurs within ten years unless the resultant severe anaemia is reversed and erythropoiesis is suppressed by regular blood transfusions. Some individuals inherit two β-thalassaemia mutations but require only intermittent transfusions and their symptoms are not severe.

Although significant psychosocial problems have been reported, this clinical syndrome arises as a result of a number of genotypes, including mild β-thalassaemia mutations, which allow some adult Hb (Hb A) production.

c) Alpha-thalassaemia

Alpha-thalassaemia major, where no α-globin is produced, is associated with intrauterine death (except when intrauterine transfusion has been undertaken) and potentially fatal maternal complications. Hb H disease occurs when three of the four α-genes are non-functional. It is of variable severity, but generally presents a thalassaemia intermediapicture (Higgs, 1993)

d) Sickle Cell Disease

The amino acid substitution in S results in polymerization / crystallization of the S molecules within the red blood cell on deoxygenating This polymerization produces a change in the cell from a biconcave disc to a crescent or sickle shape. On re-oxygenation, the red blood cell initially resumes its biconcave disc shape but, after repeated cycles of "sickling and unsickling", it is damaged permanently, becomes dehydrated and irreversibly sickled, and haemolyses (Lane, 1996)

2.6 Models of Screening

In theory, because of the inherited nature of the haemoglobinopathies, once a diagnosis has been made, only one definitive test should be necessary during one's life, as long as that information is available life-long and whenever that person comes into contact with health services. Strategies that can be adopted include preconception, antenatal, opportunistic and neonatal screening. A variety of models of haemoglobinopathy screening exist and the service is patchy and often unstructured. The varied distribution of the "at-risk" population may demand different service models, depending on prevalence, but these should be based on consideration of the benefits and outcomes of screening, the access to health care of those at risk, and issues of equity. Screening programmes may be opportunistic or systematic, targeted or population based. In selective programmes, attention needs to be given to the criteria for selection and the population base (e.g. community versus hospital).

Many people carry one haemoglobin S gene (sickle-cell trait) with no significant health problems as a result. But these carriers of the sickle-cell gene can pass the gene to their

children. When both parents are carriers, they have a one in four (1 in 4) chance with every pregnancy of having a child with sickle-cell anaemia.

2.7 Geographical Distribution of Haemoglobin S (HbS)

Certain ethnic populations have more people who are carriers of the sickle-cell trait. . The haemoglobin S gene is particularly common in western Africa and people of western African ancestry, and an estimated 8 to 12 percent of all African Americans carry the sickle-cell gene. Sickle-cell anaemia is again found in people from Mediterranean countries, the Middle East, and India, or people whose ancestors came from these regions (Eckman, 2006) There is a wide variation in the prevalence of the gene in different parts of Africa. However, the frequency of the trait has been estimated to be as high as 25-40%. In Nigeria the figure is about 25%, while the homozygous state is found in about 3% of the population (Adekile and Azubuike, 1999). Researchers believe that the haemoglobin S gene is particularly common in these populations because carriers of the sickle-cell gene are less susceptible to malaria, once one of the leading causes of illness and death in these malaria endemic regions. The sickle cell gene confers on individuals the likelihood to resist malaria thereby sustaining the S gene in the population.

2.8 Pathophysiology of Sickling

Deoxygenated HbS molecules are insoluble and polymerize. The flexibility of the cells is decreased and they become rigid and take up their characteristic sickle appearance. (These distorted cells are called sickle cells because of their resemblance to the sickle, a type of crescent-shaped cutting blade used in agriculture). The process is initially reversible but, with repeated sickling, the cells eventually lose their membrane f exibility and become irreversibly sickle. This is due dehydration (Murphy etal; 2005). Sickling can produce:

- a shortened red cell survival, leading to a deficiency of red blood cells, known as anaemia
- impaired passage of cell through the microcirculation, leading to obstruction of small vessels and tissue infarction.
- Sickling is precipitated by infection, dehydration, cold, acidosis or hypoxia.

a) Pain Crises

Pain crises in persons with sickle cell anaemia are intermittent painful episodes that are the result of inadequate blood supply to body tissues. The impaired circulation is caused by the

blockage of various blood vessels from the sickling of red blood cells. The sickled red blood cells slow or completely impede the normal flow of blood through the tissues. This leads to excruciating pain requiring hospitalization and narcotic medication for relief. The pain typically is throbbing and can change its location from one body area to another. Bone is frequently affected. Pain in the abdomen with tenderness is common and can mimic appendicitis. Fever frequently is associated with the pain crises. A pain crisis can be promoted by preceding dehydration, infection, injury, cold exposure, emotional stress, or strenuous exercise. As a prevention measure, persons with sickle cell anaemia should avoid extremes of heat and cold. The pain episodes of sickle cell disease can vary in frequency and severity. Beginning at a young age and continuing through adulthood, these pain episodes can prevent individuals from attending school and work and interfere with daily life (Claster & Vichinsky, 2003). Children and adults must be closely monitored for infection and sepsis. Infection may be marked by pain, swelling, and fever (Claster & Vichinsky, 2003). Even low-grade temperatures must be considered when evaluating an individual for infection (Claster & Vichinsky, 2003). These infections most often include Salmonella species and *Staphylococcus aureus* (Claster & Vichinsky, 2003). Human parvovirus B19 infection is responsible for approximately 80% of aplastic crises, when red blood cell production is significantly reduced in the bone marrow (Claster & Vichinsky, 2003).

The haemolysis that occurs due to the sickled shape of blood cells can lead to chronic anaemia, jaundice, and delays in growth while increased red blood cell adherence to the endothelium increases the likelihood of vaso-occlusion and organ damage. Red blood cells can become trapped in the spleen causing decreased haemoglobin concentration and enlargement of the spleen. This process called splenic sequestration can increase damage to the splenic tissue and potential for infection (Claster & Vichinsky, 2003). As sickled red blood cells accumulate in the endothelium of the cerebral arteries, there is a potential for brain injury.

b) **Fatigue and Anaemia**

Fatigue is a common symptom in persons with sickle cell anaemia. Sickle cell anaemia causes a chronic form of anaemia which can lead to fatigue. The sickled red blood cells are prone to breakage (rupture) which causes a much shorter life span of these cells (the normal life span of a red blood cell is 120 days). These sickled red blood cells are easily detected with a microscope examination of a smear of blood on a glass slide. Typically, the site of red blood cell production (bone marrow) works overtime to produce these cells rapidly,

blockage of various blood vessels from the sickling of red blood cells. The sickled red blood cells slow or completely impede the normal flow of blood through the tissues. This leads to excruciating pain requiring hospitalization and narcotic medication for relief. The pain typically is throbbing and can change its location from one body area to another. Bone is frequently affected. Pain in the abdomen with tenderness is common and can mimic appendicitis. Fever frequently is associated with the pain crises. A pain crisis can be promoted by preceding dehydration, infection, injury, cold exposure, emotional stress, or strenuous exercise. As a prevention measure, persons with sickle cell anaemia should avoid extremes of heat and cold. The pain episodes of sickle cell disease can vary in frequency and severity. Beginning at a young age and continuing through adulthood, these pain episodes can prevent individuals from attending school and work and interfere with daily life (Claster & Vichinsky, 2003). Children and adults must be closely monitored for infection and sepsis. Infection may be marked by pain, swelling, and fever (Claster & Vichinsky, 2003). Even low-grade temperatures must be considered when evaluating an individual for infection (Claster & Vichinsky, 2003). These infections most often include Salmonella species and *Staphylococcus aureus* (Claster & Vichinsky, 2003). Human parvovirus B19 infection is responsible for approximately 80% of aplastic crises, when red blood cell production is significantly reduced in the bone marrow (Claster & Vichinsky, 2003).

The haemolysis that occurs due to the sickled shape of blood cells can lead to chronic anaemia, jaundice, and delays in growth while increased red blood cell adherence to the endothelium increases the likelihood of vaso-occlusion and organ damage. Red blood cells can become trapped in the spleen causing decreased haemoglobin concentration and enlargement of the spleen. This process called splenic sequestration can increase damage to the splenic tissue and potential for infection (Claster & Vichinsky, 2003). As sickled red blood cells accumulate in the endothelium of the cerebral arteries, there is a potential for brain injury.

b) **Fatigue and Anaemia**

Fatigue is a common symptom in persons with sickle cell anaemia. Sickle cell anaemia causes a chronic form of anaemia which can lead to fatigue. The sickled red blood cells are prone to breakage (rupture) which causes a much shorter life span of these cells (the normal life span of a red blood cell is 120 days). These sickled red blood cells are easily detected with a microscope examination of a smear of blood on a glass slide. Typically, the site of red blood cell production (bone marrow) works overtime to produce these cells rapidly,

attempting to compensate for their destruction in the circulation. Occasionally, the bone marrow suddenly stops producing the red blood cells which causes a very severe form of anaemia (aplastic crises). Aplastic crises can be promoted by infections that otherwise would seem less significant, including viruses of the stomach, bowels and the flu (influenza).

The anaemia of sickle cell anaemia tends to stabilize without specific treatments. The degree of anaemia is defined by measurement of the blood haemoglobin level. Haemoglobin is the protein molecule in red blood cells which carries oxygen from the lungs to the body's tissues and returns carbon dioxide from the tissues to the lungs. Blood haemoglobin levels in persons with sickle cell anaemia are generally between 6 to 8 gm/dl (normal levels are above 11 gm/dl). Occasionally, there can be a severe drop in haemoglobin requiring a blood transfusion to correct the anaemia (such as in patients suffering splenic sequestration). Blood transfusion is usually reserved for those patients with other complications, including pneumonia, lung infarction, severe leg ulceration, or late pregnancy. (Among the risks of blood transfusion are hepatitis, infection, immune reaction, and injury to body tissues from iron overload.) Transfusions are also given to patients to prepare them for surgical procedures. Folic acid is given as a supplement.

Patients with sickle cell disease may experience acute chest syndrome, a condition characterized by fever, chest pain, and/or difficulty breathing. The appearance of a new pulmonary infiltrate on radiological studies of the chest is evidence for a diagnosis of acute chest syndrome and can be caused by infection and/or infarction. Both bacteria and viruses may be the infectious agents responsible. However, the most common cause of acute chest syndrome is a fat emboli from an infarction of the long bone which travels to the lungs (Claster &Vichinsky, 2003; Wethers, 2000).

c) Lung and Heart Injury

Aside from lung infection (pneumonia), the lungs of children with sickle cell anaemia can also be injured by inadequate circulation of blood which causes areas of tissue death. This lung damage can be difficult to distinguish from pneumonia. These localized areas of lung tissue damage are referred to as pulmonary infarcts. Pulmonary infarcts often require a special x-ray test using a dye injected into the affected areas (angiogram) for diagnosis. Repeated pulmonary infarcts can lead to scarring of the lungs of children with sickle cell anaemia by the time they reach adolescence. The heart is frequently enlarged in children with sickle cell anaemia. Rapid heart rates and murmurs are common. The heart muscle can also

be injured by infarcts and iron depositing in the muscle as it leaks from the ruptured red blood cells. Injuries to the lungs or heart are treated according to the specific type of damage and the degree of impairment of organ function. Supplementary oxygen can be required. Infections of the lungs require aggressive antibiotics. Transfusions can sometimes help prevent further damage to the lung tissue. Heart failure can require chronic heart medications to assist the heart in pumping blood to the body (Medicine net, 1996).

d) Aseptic Necrosis and Bone Infarcts

Inadequate circulation of the blood, which is characteristic of sickle cell anaemia, also causes areas of death of bone tissue (bone infarction). Aseptic necrosis, or localized bone death, is a result of inadequate oxygen supply to the bone. Aseptic necrosis is also referred to as osteonecrosis. While virtually any bone can be affected, the most common are the bones of the thighs, legs, and arms. The result can permanently damage or deform the hips, shoulders, or knees. Pain, tenderness, and disability frequently are signs of aseptic necrosis. Painful bone infarcts can be relieved by rest and pain medications.

Aseptic necrosis can permanently damage large joints (such as the hips or shoulders). Local pain can be relieved and worsening of the condition can be prevented by avoiding weight bearing. With more severe damage, total joint replacement may be needed to restore function.

e) Eye Damage

The critical area of the eye that normally senses light is called the retina. The retina is in the back of the eye and is nourished by many tiny blood vessels. Impairment of the circulation from the sickling of red blood cells results in damage to the retina (retinopathy). The result can be partial or complete blindness.

Bleeding can also occur within the eye (retinal hemorrhage) and retinal detachment can result which in turn lead to retinal detachment can lead to blindness.

Once blindness occurs, it is usually permanent. Preventative measures, such as laser treatments, can be used if bleeding into the eye and retinal detachment are detected early.

f) Other Features

Additional features of sickle cell anaemia include weakening of bones from osteoporosis, kidney damage and infection, and nervous system damage. Osteoporosis can lead to severe pain in the back and deformity from collapse of the bony building blocks (vertebrae) of the spine. Kidney damage can lead to poor kidney function with a resulting imbalance of blood sodium and acidity as well as bleeding into the urine. Kidney infection can cause pelvic pain

and require hospitalization with antibiotic treatment. Injury to the nervous system can result from meningitis or sickle cell anaemia itself. Poor blood circulation in the brain can cause stroke, convulsions, and coma. Damage to the brain from stroke can cause permanent loss of function to areas of the body. Transfusion of blood and fluids intravenously can be critical. Medications to reduce the chance of seizures are sometimes added. If stroke results in long-term impairment of function, physical therapy, speech therapy, and occupational therapy can be helpful.

Individuals with sickle cell disease have a shortened lifespan in comparison to their healthy counterparts. However, cohort studies following children with sickle cell disease from birth to age 18 years have demonstrated an increase in the mean age at death and a decrease in childhood mortality. Infection has been identified as the number one cause of death in sickle cell patients, but studies are demonstrating a decrease in mortality rates due to infection. This increase in survival and decrease in mortality due to infection are attributed to early identification of disease through newborn screening and early intervention through prophylactic antibiotics (Quinn et al, 2004).

2.9 Management of Sickle Cell Disorder

There is no universal cure for SCD and treatment options are rather limited, however improved knowledge has greatly advanced medical management over the past four decades. Antibiotic prophylaxis is used to prevent infections especially in children. Other therapies aim to minimise the effects of symptoms of the disorder. Painful episodes (crises) are managed primarily with analgesia, and hydration. Analgesic pain control is usually in progressive stages and requires a variety of medications ranging from paracetamol for mild pain to morphine for severe pain. Blood transfusions may be required for stroke and other complications, and hydroxyurea has also been found to be very effective in reducing the 'sickling' process and consequently the frequency of pain and hospitalisations experienced by patients. Bone marrow transplantation is a possible cure; however among other criteria, this requires a matched donor, and unfortunately is not feasible for all affected children.

2.10 Diagnosis

Screening tests

These are tests that indicate the presence of HbS, but do not define the Hb genotype of the individual. The tests are based either on the morphological changes that occur in red blood

cells containing HbS, when subjected to deoxygenating (sickling test), or on the poor solubility of HbS in solutions of high molarities (solubility test).

Haemoglobin Electrophoresis

Sickle-cell anaemia is diagnosed by a procedure called haemoglobin electrophoresis, in which haemoglobin samples are identified by the speed of the haemoglobin when subjected to an electric field. Several different media, supporting structures and buffers have been used, but the most widely used is cellulose acetate electrophoresis (buffer pH 8.6). It is fast and generally reliable.

Citrate agar electrophoresis (pH 6.3) is an alternative to cellulose acetate and should be used as a confirmatory test for suspicious patterns obtained with the laner. It gives good separation of the common Hb variants (Adekile, 1999).

2.11 Public Health Importance of Sickle Cell Disorder

Sickle Cell Disease (SCD) and Thalassaemia are classified as the two main Haemoglobin Disorders, and in recent years have been acknowledged to have a global impact by the World Health Organisation (WHO). SCD comprises a group of inherited red blood cell conditions that result from the synthesis of variant or mutant haemoglobins. Over 300,000 babies are born worldwide with SCD mostly in low and middle income countries, with the majority of these births in Africa (Fleming and Watkins, 2005). SCD originates in tropical regions as a result of its advantage against malaria. It is predominant among people from African, Asian, Arabian and Mediterranean countries; nonetheless it is a global health problem because of population migration. SCD results in early childhood death if left untreated, and its effect on the burden of health care is being recognised as a global issue in terms of chronic disease. Inheritance of a single sickle haemoglobin (HbS) gene results in a healthy sickle cell carrier state, while the inheritance of the HbS gene from both parents, or HbS with another variant haemoglobin gene (e.g HbC, Hbβ-thalassaemia) results in symptomatic SCD.

Generally, the prevalence of healthy carriers (sickle cell trait) ranges between 10% and 40% across equatorial Africa and decreases to between 1% and 2% in north-eastern Africa and less than 1% in Southern Africa. In West African countries such as Ghana and Nigeria, the frequency of carriers is 15% to 30% while in East African countries such as Uganda and Tanzania it shows wide variations of up to 45% in some areas (Adeyemo etal; 2007). This distribution is thought to reflect current or historic exposure to plasmodium malaria infection

as carriers appear to be protected from malaria associated deaths and thus have improved survival and therefore continued transmission of the HbS gene. The incidence of SCD at birth is determined by the prevalence of carriers in the population. SCD has remarkable public health implications for Africa. It contributes the equivalent of 5% to under-five deaths in Africa, with up to 16% in West Africa. Thus in Nigeria, with an estimated carrier prevalence of 24%, 20 per 1000 births are estimated to be affected by SCD, resulting in 150,000 children with SCD born annually in Nigeria.

The trauma experienced from sickle cell disease cannot be over emphasized. Katibi (2008) stated that patients with sickle cell disease may have recurrent illness and be hospitalized due to various complications of the disease. The cost implication and mental agony of the parents in particular are of significant note. He further identified physical deformities of the patients such as frontal bossing, protruding abdomen, thin extremities and gnathopathy.

2.12 Knowledge and Perception of Sickle Cell among African-Americans

A limited number of studies have been conducted to examine African-Americans' knowledge and perception of sickle cell disease, sickle cell trait, carrier detection, and newborn screening. Past research has demonstrated a significant lack of awareness regarding the difference between disease and trait status and how trait status increases the chance to have a child with disease (Midence, 1994; Treadwell, 2006; Wright, 1994). Specifically, one study of 147 African-Americans between 18-50 years of age found that 31% knew if they were carriers for sickle cell trait (Wright, 1994). A majority (73%) did identify that sickle cell is a genetic disease (Wright, 1994). Misconceptions of how one inherits sickle cell disease have also been documented among the African-American community. Interestingly, a study of individuals with sickle cell disease found that 23% believed sickle cell trait could turn into sickle cell disease (Midence, 1994). A 2006 study was undertaken to look more closely at African-American community members' knowledge and misconceptions of sickle cell (Treadwell, 2006). Three focus groups and 282 surveys were conducted in a metropolitan area of California (Treadwell, 2006). Focus group participants were asked to identify barriers to follow-up counselling for sickle cell trait detected by newborn screening, to describe their understanding of sickle cell disease, and to suggest possible solutions for poor follow-up rates for sickle cell trait counselling (Treadwell, 2006). Common themes among community members who participated in these focus groups included limited visibility of sickle cell disease and trait and the need to use media to promote awareness (Treadwell, 2006).

participants discussed the stigma associated with any type of disease in the African-American community and the need for health care professionals to have compassion and cultural sensitivity when discussing sickle cell (Treadwell, 2006).

Surveys were designed to determine where individuals receive their information about sickle cell, what individuals currently know about sickle cell disease and trait, if individuals know their trait status, and the effectiveness of different sources of information in improving knowledge about sickle cell (Treadwell, 2006). Among respondents, 86% could identify that sickle cell causes serious health problems, and 91% stated that sickle cell disease is most prevalent among African-Americans (Treadwell, 2006). Approximately 86% of respondents knew that sickle cell disease is inherited from both parents, and slightly fewer respondents were able to correctly identify the reproductive risks of a sickle cell trait carrier (Treadwell, 2006). Eighty-one percent of individuals believed if you have sickle cell trait you have a chance to have a child with disease, and 78% of individuals believed a child with sickle cell trait would be at risk to have a child with disease in the future (Treadwell. 2006). Of survey participants, only 15.9% knew their trait status and of those individuals, 53% learned their trait status through discussion with family members (Treadwell, 2006). The greatest majority of individuals receive their information about sickle cell disease and trait from friends and acquaintances (Treadwell, 2006). Respondents who received information from friends and family were three times more likely to know their trait status (WHO, 2006). These findings emphasize the benefit of family discussion about sickle cell and the risk within a family.

2.13 Attitudes toward Genetic Testing

The high degree of perceived benefit from genetic testing and counselling for SCD identified by Gustafson and colleagues (2007) is of significance given that several research studies examining the general attitudes and beliefs of African-Americans toward genetic testing have found participants hold both serious concerns and positive beliefs about the benefits of testing. Studies conducted among individuals in clinical, educational, and community settings consistently demonstrate that minority participants describe considerable risks associated with genetic testing including, but not limited to, the possible misuse of genetic information for the purpose of racial discrimination. However, minorities also describe benefits of genetic testing for prevention of disease (Catz et al. 2005; Kessler et al. 2005; Laskey et al. 2003; Zimmerman et al. 2006).

In contrast to the study by Catz and colleagues (2005) which examined attitudes toward genetic testing among minorities with a range of educational levels, Laskey and colleagues (2003) pursued similar questioning among African-American college students in a premedical program. These undergraduates also stated that genetic testing could benefit individuals/families by allowing for prevention or preparation. However, the group's overall concerns about the risks of genetic testing were more theoretical in comparison to the responses obtained by Catz and colleagues. Concerns about genetic testing included risk of genetic discrimination, increasing abortion rates, eugenics, and breach of confidentiality.

2.14 Genetic Testing and Counselling Services in Nigeria

The diversity and heterogeneous distribution of haemoglobin disorders make it necessary to develop strategies at the country and state level. WHO has recommended global development of these services (WHA, 2006). However, service development can be unexpectedly challenging, because it requires inclusion of genetic approaches in health systems. This is the main reason why there are scanty or few numbers of designated centres for sickle cell testing and counselling services. Though there are no designated centres for about to wed couples for premarital screening in Nigeria per se, but almost all the medical facilities with laboratory offer testing and counselling services on demand by patients. Therefore, the service is open to any person. There are also some denominations that compel about to wed couples who are their members to do their testing and counselling in their facilities.

Sickle cell foundation of Nigeria is an organization that has a designated package for genetic testing and counselling services. They deliver genetic counselling at the National Sickle Cell Centre, Sickle Cell Clinics and Sickle Cell Clubs with trained personnel – volunteer or paid staff (SFN, 2011)

2.15 Theoretical Framework

Researchers investigated models related to attitudinal and behavioural change. Health Belief Model (HBM) is employed for this study.

The model is a psychological model that attempts to explain and predict health behaviours. It focuses on attitudes and a belief of individuals. The model explains and predicts health behaviour with special reference to preventive health behaviours by focusing on the perceptions and beliefs of individuals. The HBM was first developed in the 1950s by social psychologists Hochbaum, Rosenstock and Kegels working in the U.S. Public Health Services (National Cancer Institute, 2005). The model was developed in response to the failure of a

free tuberculosis (TB) health screening program. Since then, the HBM has been adopted to explore a variety of long- and short-term health behaviours, including sexual risk behaviours and the transmission of HIV/AIDS.

Perceived susceptibility – measures individual's perception of his risk of possibility of having and transmitting genetic disorder trait (Sickle cell trait carrier parent). The combination of perceived susceptibility and perceived severity is considered a threat. Therefore, in this model, if intending couples could be aware of the perceived susceptibility and severity of SCD they would initiate a positive health behaviour that would prevent incidence of SCD.

Perceived severity - perception of potential seriousness of the consequences of disorder if transmitted (existence of having a child with sickle cell disorder)

Perceived barriers - refer to the negative factors discouraging behavioural change, such as cost, confidentiality cum privacy, denial of benefits e.t.c. In this case they represent factors that may that hinder intending couples from utilising PSCCT service.

Perceived benefits – These include positive aspects of engaging in a behaviour that have to outweigh the negative aspects such as convenience, time, money, improved knowledge and awareness that will help in the utilization of premarital genetic testing and counselling.

Self-Efficacy -this is perceived control i.e. the self-confidence an about to wed spouse have to utilize pre-marital genetic screening will encourage him/her to go for it. Self- efficacy in this case is the display of potentials that one can engage ina health promoting behaviour.

Cue to Action – refers to strategies that are in place to ginger about to wed spouses to take premarital genetic testing and counselling. Intending couples may require external factors such as advice from close relatives, a physician, and daily reminder such as mass media, documentaries. Significant others like marriage counsellor, mentors, death of a family member who had sickle cell disorder may also influence intending couples to take action.

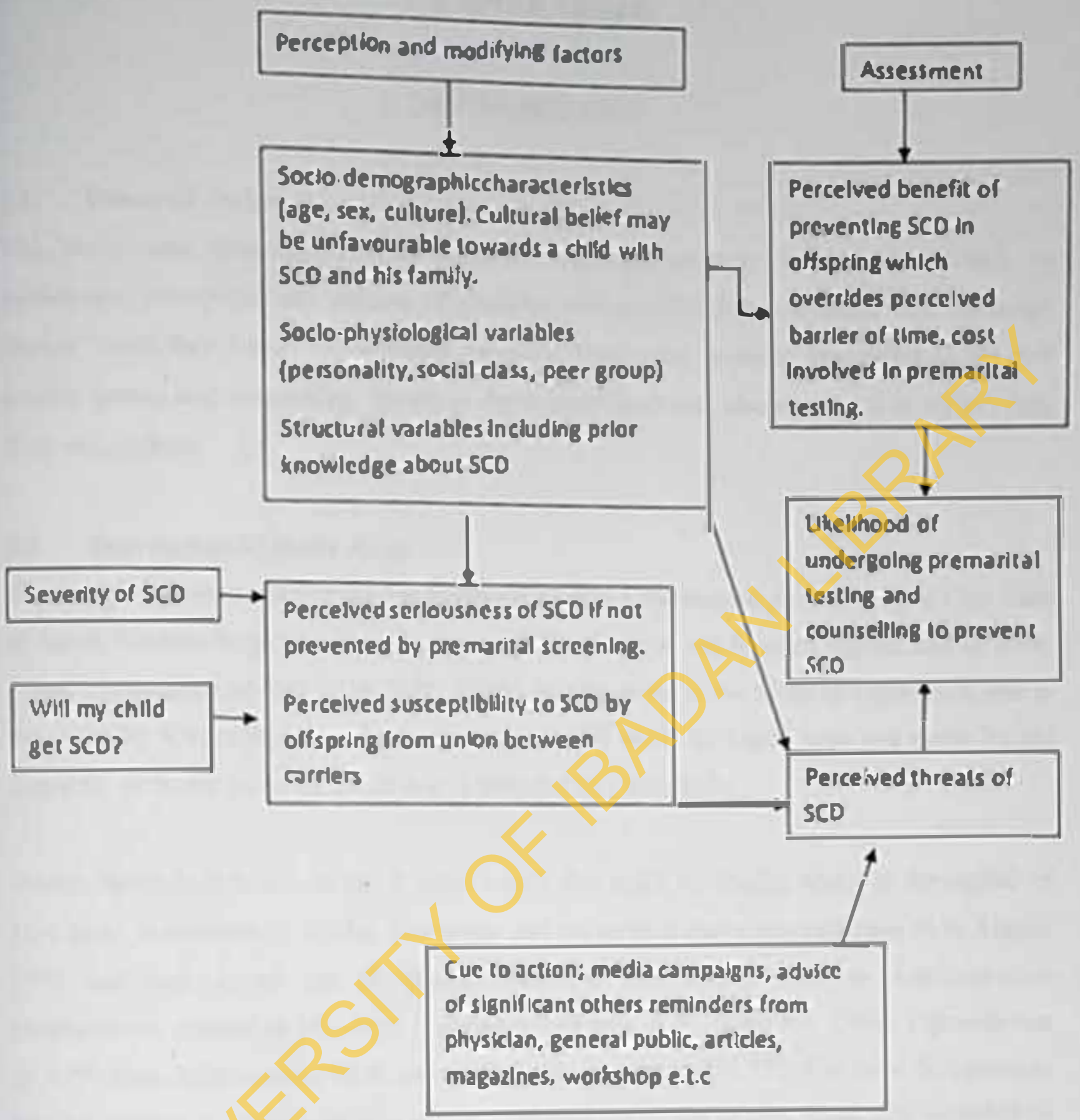


Fig 2.1 Application of Health Belief Model to Utilisation of PSCT

CHAPTER THREE

METHODOLOGY

3.1 Research design

The study was descriptive, cross-sectional local government based and focused on knowledge, perception and attitude of about-to-wed couples that contracted their marriage Ibadan North-East Local Government Registry, Oyo state, towards premarital sickle cell genetic testing and counselling. Seven In-depth interviews was also employed to collect data from respondents

3.2 Description of Study Area

The study was carried out in Ibadan North-East Local Government Area (LGA) of Oyo State in South Western Nigeria. It was created in 1976 out of the old Western region; and in 2006, it had a population of 6,617,720 (NPC, 2006). It is bounded in the south by Ogun State and in the north by Kwara State; on the west, it is bounded partly by Ogun State and partly by the Republic of Benin while on the east, it is bounded by Osun State.

Ibadan North-East is one of the 5 urban LGAs that make up Ibadan which is the capital of Oyo State. It contains 11 LGAs; five urban and six rural. It came into existence in in August 1991 and was carved out of Ibadan Municipal Government with its Administrative Headquarters situated at Idi –Ape. It covers a land area of 51.250sq km. Using a growth rate of 3.2% from 2006 census, 2010 estimated figure was put at 374,772. The local Government has the highest population density of 7,313 persons per sq km in Oyo State. It is bounded by Ona-ara, Ibadan South-East and Egbeda Local Government Areas. Ibadan North-East Local is an urban centre which forms part of Ibadan Metropolis. The residents are Yoruba and other tribes notable for banking and trading activities. A considerable number of commercial banks are lined up among major streets of the Local Government Area. It has the largest spare part markets called Araromi market, gate (Iso part).

Ibadan NorthEast is subdivided into 12 wards.

3.3 Population of Study

Population of study consisted of about-to-wed couples who contracted their marriage in Ibadan North-East Local Government secretariat marriage registry, Oyo State.

3.4 Inclusion Criteria

- Only the registered respondents who were obviously present at the registry.
- Respondents must conduct their marriage in Ibadan North East marriage registry during the period of data collection.
- Those who came for registration to announce their intentions and consented to participate in the study.

3.5 Exclusion Criteria

- About-to-wed respondents that were not sighted at the time of research were excluded.

3.6 Determination of Sample Size

The sample size was obtained by the use of Epi info statistical software.

$N = Z^2 p q/d^2$ (Leslie Kish, 1965)

Where N = minimum sample size

Z is a constant put at 1.96 at 95% confidence interval

P = prevalence

Q = 1 = p

D = 5% desired precision (degree of accuracy)

The design effect is 1

P = Assumed prevalence 50%

P=50%

Q= 1 – 0.50 = 0.50

N = [(1.96) ^2 × 0.5×0.5)/ 0.05^2

= $((1.96)^2 \times .5(.5)) / (.05)^2$

= (3.8416 x .25) / .0025

= 0.9604 / 0.0025

= 384

385 respondents are needed

However, the sample size (N) was increased to 400 in order to increase precision in variables measured.

3.7 Sampling Technique

The study was conducted to investigate level of knowledge, perception and attitude of about-to-wed couples towards premarital sickle cell counselling and testing in Ibadan North-East

Local Government Area marriage registry in Oyo State. Purposive sampling was used to select 400 consenting respondents.

3.8 Variables

The variables were categorised into two namely the independent and the dependent variables.

3.8.1 Independent Variables

Independent variables are socio-demographic characteristics of respondents, level of education, religion, current activity, living status and age.

3.8.2 Dependent Variables

The dependent variables in the study are knowledge, perception, attitude and utilization of about-to-wed couples towards Premarital Sickle Cell Genetic Testing and Counselling in Ibadan North East Local Government Registry, Oyo state.

3.9 Methods and Instruments for Data Collection

A mixed method was used to collect data consisting of In-depth interview (IDI) guide and a questionnaire.

Instrument for Data Collection

(i)In-depth Interview

In-depth interview (IDI) constitutes 10 questions (Appendix. for the IDI).The questions were framed and tailored to give further insights into knowledge, perception and attitude of about-to-wed couples towards premarital sickle cell counselling and testing in Ibadan North-East Local Government Area, Oyo State was used to facilitate the discussion

(ii) Questionnaire

The collation of quantitative data was done by means of semi-structured questionnaire (Appendix). The questionnaire was developed after a review of the literature. The questionnaire used to elicit quantitative data from the respondents consists of five sections labelled I – V. Section I sought information on Age, level of education, Religion, Ethnicity, Current activity, Living status and Level of Education. Section II contained questions relating to knowledge about Sickle Cell Disorder. Section III sought information on utilisation of Premarital Sickle Cell Counselling and Testing services Section IV focused on eliciting information on Premarital Sickle Cell Counselling and Testing. V focused on eliciting information on perception of sickle cell disorder and health beliefs.

3.10 Validity of the Instrument

To ensure validity of the instrument (the questionnaire and IDI guide) for data collection, the following steps were taken;

The drafts of questionnaires were developed by consulting relevant literature. The draft of the instrument went through an independent review from peers, my supervisor and expert researchers in the Faculty of Public Health, College of Medicine, and University of Ibadan. The experienced researchers consisted of specialists in Health Promotion, Education, Population and Reproductive Health. Pre-test of the instrument was conducted between April and May, 2014 using Ibadan North LGA marriage registry based on similarities in characteristic with the study population. Supervisor review was used in fine-tuning the instrument. Content validity of the questionnaire was further ensured through the incorporation of the preliminary pretested IDI outputs. Special care was taken to monitor the quality of data collected through supervision during data collection. Forty questionnaires were administered in Ibadan North LGA marriage registry. The completed questionnaires were edited, responses coded and entered into a computer. The data were then analyzed using descriptive and inferential statistics.

Based on the pre-test results, the following recommendations were made;

- The title of the study was modified to read "knowledge, perception and attitude of about-to-wed couples towards premarital sickle cell counselling and testing in Ibadan North-East Local Government Area"
- The questionnaire was reduced to 5-page and the average administration time per Questionnaire was recorded as 30minutes.

3.11 Reliability of the Instruments

To ensure reliability of the instruments (the questionnaire and IDI guide) were reviewed for quality and consistency. The two instruments were pre-tested to ascertain sustainability and appropriateness to field situation, determine whether the questions were clear and concise enough for participants comprehension and determine the trend in the response of participants and the amount of time it took to administer the questionnaire. The two instruments used for data collection were pre-tested to ascertain suitability and appropriateness to field situations, determine whether the questions were clear and simple enough for participants' comprehension and determine the trend in the response of participants and the amount of time it took to administer the questionnaire.

The IDI guide was pre-tested among about to wed couples in Ibadan North LGA Oyo State registry. Forty respondents were interviewed using the questionnaire (representing 10% of the actual sample size for this study) at Ibadan North, LGA marriage registry. The questionnaires were cleaned, coded and entered into the computer. The reliability of the questionnaire was

determined using the Cronbach's Alpha model technique of SPSS (version 15). The reliability correlation co-efficient of 0.76 was obtained, which indicated that the instrument was reliable. The outcome of the pre-test was used to correct and modify questions which seemed repeated, unclear to respondent and those that were found to be irrelevant were removed and adequate spaces were provided for responses as well as skipping mechanism.

3.11.1 Recruitment and Training of Research Assistants for the study

Despites the fact that the research, there was still need to recruit and train Research Assistants (RAs) who would help in data collection. A 4-man team of researchers comprising of the principal investigator was constituted. The following selection criteria were employed to select the four (4) RAs for training with the aim of selecting three (3) thereafter.

- Educational qualifications of the assistants were at least Ordinary National Diploma (OND), BSc in a health and/or science related field, MSc and MPH.
- The candidates were fluent in English.
- Interpersonal and good communication skills.
- Report writing skills.
- Ability to devote all hours to the research work while it lasts.

3.11.2 Training of Research Assistants

The research assistants were trained for two days 20th to 21st March, 2014. A training manual, plan and timetable were developed and approved by the project supervisor for the training. A time table was drawn for this period of 3 hours 9a.m-12 noon daily at the students' lounge of the Faculty of Public Health, University College Hospital. The training commenced with introduction of the trainer or the principal investigator and trainees. The trainees received training materials, sessions introduced them to the research study, objectives and methodology, role-plays on the data collection procedure (entry processes, seeking consent of potential respondents for the study, signing of confidentiality assurance form and administration of questionnaire). The appropriate training methods and materials for facilitation were selected. These methods included a combination of largely active training methods such as participatory discussions, demonstration and return demonstrations and lectures to make the training participatory. Review questions for monitoring and assessing trainees' comprehension was asked intermittently.

Demonstrations were used to transfer skills for conducting IDI, Transcriptions and report writing of findings, and especially for the correct interpretation of each item. The questionnaire and the IDI guide was revised with them during the training after which the RAs were equipped with copy of the instrument each to be taken home and read over for better understanding with aim of answering any burning question that may result the following day. The questionnaire was further reviewed to ensure the consistency in the numbering and the adequacy of the skip instructions. In addition, content and construct validity were reviewed during the training of research assistants to ensure uniform understanding and interpretation by all research assistants. Negotiations and logistic plans for data collection were discussed and stipends paid to RAs. The researcher facilitated the training with supervision by colleagues who are experienced and have previously worked as research assistants. This team helped to assess and score trainees and based on the assessment scores, the final three research assistants were selected for the study. Each RA was assigned potential dates for data collection and were directly supervised by the researcher. Each RA received a copy of the field manual, copies of the questionnaire, one copy of the ethical approval from the State Ministry of Health and writing materials all contained in a clear water proof bag. All RAs participated in the data collection for the pre-test of the questionnaire in Ibadan North LGA marriage registry.

3.12 Data Collection Procedure

The study was carried out from June to Dec 2014 with the assistance of the three trained Research Assistants. The principal researcher with the four trained research assistants administered the questionnaires to the respondents in Ibadan North-East local Government Area marriage registry. The research assistants were trained in the following areas; the objectives of the study, the sampling procedure, how to secure respondents informed consent; importance of collecting valid data; procedures for questionnaire administration and techniques for reviewing questionnaires for reviewing of the items on the questionnaire to have adequate understanding of the instrument and completeness. The manual of field operation was prepared to explain how entries would be made, the number of questionnaires to be administered and how variables would be coded. The research assistants with the researcher were involved in the collection of the data. Data collection took place in the month of June to early December, 2014 mostly in the morning on week days. Short debriefing sessions were also held at the end of each day where the day's work was reviewed and the next plan of action conveyed to the research assistants.

3.12.1 Qualitative Method

A total of seven IDIs were conducted among intending couples in early June, 2014 for two days. The sessions were conducted in a venue that was free of distraction. The venues were carefully selected with input of craft masters. The venues used were conducive for the participants to discuss freely. Each IDI session lasted for a minimum duration of 45 minutes. The discussion sessions were conducted by a moderator, one note taker, who documented discussants responses using a tape recorder and through verbatim writing of proceedings. The discussants were provided with full details of the study and an assurance of confidentiality of the disclosed information. Permission to use a tape recorder was sought and verbal consent was obtained from discussants prior to commencement of each session. Each discussion was a highly interactive as discussants took turns to air their views in respect of the issues raised for discussion. Discussants were encouraged to feel free to express themselves. Each of the IDI was later transcribed and the information used to support the report generated by the note-takers.

3.12.2 Quantitative Method

The quantitative data was collected with the use of a semi-structured questionnaire administered in secluded places such as; inside the shops, under the tree and veranda to ensure convenience and privacy of the respondents. The data collection process included the following steps; visit to the over-all head of the registry to seek permission to conduct interviews and administer questionnaire on the respondents. The semi-structured questionnaire was interviewer administered since majority of the respondents were literates. The sections in the questionnaire included respondents socio-demographic characteristics, while other section contains information on variables of the study.

3.13 Data Management and Analysis

The tape-recorded response from the IDI were transcribed verbatim and used to update the write up of the recorder. The IDI report was analyzed manually by the researcher. Content and context analysis using a thematic approach involving the grouping together of similar themes in each transcript which was followed by identifying emerging trends and differences across transcript.

In respect to data analysis from the questionnaire the following were done:

All the adininistered questionnaires were checked one by one and edited for the purpose of completeness and accuracy. Questionnaires were serialised for easy identification and recall of any problematic instrument and for correct data entry and analysis. A coding scheme guide was developed after carefully reviewing the responses and appropriate scoring was done. The data was inanually coded and entered into the computer for analysis. Respondents' knowledge on sickle cell disorder were measured using knowledge scale. Seven knowledge questions were asked and points allotted to each of the knowledge questions (3 points). Responses that were very close to the most correct answer were allotted 2 points each. Any response that was incorrect carried no point (i.e zero point). The total knowledge score and the maximum obtainable score for each respondent was calculated. The Knowledge scores 10, 10- 20 and > 20 were categorised as poor, fair and good respectively. Perception towards PSCCT was measured on a 20-point scale; scores 10 and >10 were classified as negative and positive respectively. Attitude was measured on a 20-point scale on which scores 10 and >10 were classified as negative and positive respectively. The data were analyzed using IBM Statistic Package for Social Science (SPSS) (Version 16) statistical package Micro soft excel version 2007 for windows were used in the data analysis. The descriptive statistical tools used were mean, standard deviation and the inferential statistics of Chi-square (X^2) and logistic regression test were used for the analysis. Frequency and percentage tables were generated and Cross tabulations of some variables done using Chi-square (X^2) test. The research hypotheses were tested to establish associations between the independent and dependent variables using the Chi-square test at 5% probability level for rejecting the null hypotheses. Cross tabulation of dependent and independent variable was also done to establish associations between the variable. The results were summarized and presented in chapter four of this dissertation. In addition, narratives statements were further used to present the data.

3.14 Ethical Considerations

The proposal was submitted for approval and reviewed by the Oyo State Ethical Review Committee. Informed consent was obtained from the respondents by giving them an informed consent form to fill by explaining it to the best of their understanding. The inform consent form spelt out the title of the study, purpose of the study, justifications for doing the study as well as the benefit that will be derived at end of the study. The respondent that cannot read or write verbal information consent was sort from the respondents before the commencement of the filling of the questionnaire.

participation was voluntary and there was no criticism of respondents who refuses to participate.

Participants' identities like name or address was not written on the questionnaire so as to keep the information given by each respondent as confidential as possible. However, participants' were given equal opportunities to withdraw their consent freely during the study. Confidentiality of each participant was maximally maintained during and after the collection of his information. Information gathered from the respondent was stored in a computer system for analysis by the researcher while the questionnaire filled by the respondent was kept for maximum of ten years after which it is believed that the purpose of the study would have been accomplished

3.15 Limitations of the Study

- One of the limitations encountered during this study was getting consenting respondents.
- Another limitation was that some of respondents were not willing to divulge vital information they felt was private or personal and which were crucial to the relevance of this study.
- The researcher minimize these limitations by creating a very relaxed atmosphere that will enhance detailed recall of relevant experiences, not being discriminatory or judgmental and assuring them of confidentiality in order to encourage their total cooperation.

CHAPTER FOUR

RESULT

Basically, the findings of this study are presented in this chapter; additionally the qualitative data obtained from the IDI is also included in the results.

The results of this study are presented in this chapter. It consists of socio-demographic characteristics; Information presented in this section is intended to facilitate the interpretation of key variables relating to the knowledge, perception and attitude of about-to-wed couples towards Premarital Sickle Cell Genetic Testing and Counselling.

4.1 Socio-Demographic Characteristics

The socio-demographic characteristics are presented in Table (4.1). The respondents' age ranged from 20 – 30 years and above with a mean age of 31.73 ±5.508. A large proportion (34.5%) was aged 26 - 30 years while (33.5%) were above 30 years of age. It is obvious that, respondents within the 20 – 25 years age bracket constituted 31.8%, 55.0% were female while 45.0%were male. With regards to highest level of education, respondents with tertiary education (79.0%) topped the list, followed by those with secondary education (16.8%), primary education (2.2%), and no formal education (2.0%). Slightly above half (52.0%) of the respondents were Christians, followed by adherents of Islam (47.0%) and (1.0%) were traditionalists. Majority (66.2%) of the respondents were Yoruba, few were Igbo (20.0%) and 6.7% were Hausa. The other ethnic groups are highlighted in Table (4.1).

Most (39.8%) of the respondents were living with parents, (22.5%) were living alone, 20.8% were cohabiting, while 17.0% were living with other people.

Table 4.1 also presents detailed information about main current activity of the respondents, (38.0%) of the respondents were in paid employment, almost same proportion 30.8% were self-employed, 15.8% were schooling, 11.8% were seeking for job, 1.8% were doing housework and 0.8% were retired.

Table 4.1 Socio-Demographic Characteristics of Respondents

	N	(%)
Age		
20 – 25years	128	32.0
26 - 30years	138	34.5
>30 years	134	33.5
Total	400	100%
Gender		
Male	179	45.0
Female	221	55.0
Total	400	100%
Current activity		
Retired	3	0.8%
Paid employment	152	38.8%
Self employment	123	30.8%
House work	7	1.8%
Schooling	63	15.8%
Job seeking	47	11.8%
Total	400	100%
Highest Education Level		
No formal education	8	2.0%
Primary education	9	2.2%
Secondary education	67	16.8%
Tertiary education	316	79.0%
Total	400	100%
Religion		
Christianity	208	52.0%
Islam	188	47.0%
Traditional	4	1.0%
Total	400	100%

Table 4.1 continued

Variables	Socio-demographic characteristics of the respondents	
Ethnicity	N	(%)
Yoruba	265	66.2%
Igbo	80	20.0%
Hausa	26	6.6%
Edo	25	5.5%
Middle belt ethnic minorities*	4	1.2%
Total	400	100%
Living status		
Living alone	90	22.5%
Cohabiting	83	20.8%
Living with parents	159	39.8%
Living with others	68	17.0%
Total	400	100%

Ethnic minorities includes kogi and Benue

4.2 Knowledge of Respondents about SCD

This section presents respondents' knowledge relating to SCD and it is highlighted in Table 4.2

Majority (76.0%) of the respondents were of the view that SCD is caused by inherited sickle cell traits from both parents, only 4.8% chose none of the above. Few of the respondents (9.8%) were of the view that the bad blood is the cause of SCD, while a few (9.5%) of the respondent said virus causes it.

The table also reflects the respondents response about effects of SCD, below half of the respondents (38.8%) said SCD could cause severe debilitating pain, (10.8%) were of the view that SCD could cause organ damage, 6.0% said strokes, 7.0% chose infections while 37.5% believed that SCD could cause all the mentioned ailments.

Though, the IDI discussants could not give a scientific definition of SCD, but majority were aware of the cause of SCD and as such gave their own understanding of the health condition. Typical responses include:

- *SCD is a serious hereditary health condition which causes regular illness in the patient. It occurs in children whose parents are carriers of sickle cell traits.*
- *I cannot really give a scientific description of SCD, but I know it is a disease that causes the patient to look pale and feel sharp pains around the joint most especially during the cold weather. Sometimes, patients have blisters on the lips and sometimes have protruding abdomen.*
- *Sometimes, the patients grow lean and their eyes become yellowish and they look sick, thus have to be on an expensive and constant medication for survival, in fact it takes almost a lifetime to manage SCD.*

A very few respondents (1.0%) were of the opinion that SCD occurs in white people only, a few (1.2%) respondents also said that SCD occurs in boys only, (1.5%) also said it occurs girls only. Six percent of the respondents were of the view that only black people have SCD (6.8%), while the majority chose that SCD occurs in all categories of people mentioned.

Twenty-eight percent of the respondent believed that SCD make red cell soft and sickle (28.0%), while (27.8%) were of the view that SCD causes red cell to be to be hard and sickle

shaped. 26.5% said SCD makes red cells stiff and sickle shaped 9.5% chose round and soft, while 8.2% SCD makes red cells sticky and appear blue.

Thirty - four percent of the respondents were of the view that SCD could be managed with the use of pain relievers (34.2%), while a few (13.2%) chose blood transfusion. A few (15.8%) also considered liver transplant a management option for SCD and (15.2%) chose antibiotics. while 21.5% considered none of the options.

The IDI discussants gave different management options for managing SCD. Their responses include the following

- *SCD patients require a lot of analgesics to relieve them from agonizing pains they experience during their crisis, some of the respondents commented.*
- *Just like an HIV patient, a SCD patient has to live a healthy life by using his medications as prescribed by the physician according to some respondents.*
- *Some respondents mentioned blood transfusion and the use of antibiotics as the most popular treatment options for SCD.*

Minority (1.2%) of the respondents did not consider any cure option for SCD, (13.2%) said rest and 20.8% were of the view that antibiotics could be used. (28.5%) of the respondents agreed that bone marrow transplants option is the appropriate curative for SCD, while the majority (36.2%) chose all the listed options.

Majority (63.8%) of the respondents were of the view that diagnosis of SCD is by simple blood test. 31.5% said that they look sick, a very few (1.8%) and (3.0%) were of the view that there is no way of knowing.

Two percent of the respondents were of the opinion that condom use can prevent SCD (2.8%), while a substantial proportion (78.5%) were of the view that premarital genetic testing and counselling could prevent SCD, (2.8%) chose abortion, (3.5%) opted for all the listed options, and (12.8%) disagreed with all the options.

Fifteen percent of the respondents were of the opinion that one should test for SCD (15.0%)only when feeling sick, the majority (43.0%) said at any time, while 23.0% felt the test should be done when one is about to wed and 19.0% chose all the listed options. Twenty-eight percent of the respondents were of the opinion that the purpose of PSCCT is to make an

informed marital choice (28.8%), while 28.2% were of the view that it is to know the Sickle Cell (SC) trait status, 24.0% agreed that it is to prevent the incidence of SCD.18% of the respondents chose all the listed options, while a negligible (1.0%) chose none of the listed options.

Table 4.3 also shows the summary of knowledge of intending couples about sickle cell disease. Knowledge scale was used to assess the level of knowledge of the respondents about SCD. Respondent who scored 10, were rated as having poor knowledge. Those who 10 -20 were rated as fair in knowledge and respondents with >20 were rated as having good knowledge. Majority of the respondents (63.0%) had fair knowledge of SCD in comparison to those who were rated good (21.0%) and poor (16.0%) in knowledge SCD. In conclusion, the mean knowledge score of respondents was 11.4 ± 5.0 and 63.0% of them had fair knowledge, while 21.0% had good knowledge as shown in (Table 4.3)

Table 4.2 Knowledge about Sickle Cell Disorder

Variables	N	%
Causes of SCD		
Virus	38	9.5%
Inherited Sickle cell traits from both parents*	304	76.0%
Bad blood	39	9.8%
None of the above	19	4.8%
Total	400	100%
Effects of SCD on patients		
Severe debilitating pain	155	38.8%
Strokes	24	6.0%
Infections	28	7.0%
Organ damage	43	10.8%
All of the above*	150	37.5%
Total	400	100%
Occurrence of SCD		
Boys only	5	1.2%
Girls only	6	1.5%
White people only	4	1.0%
Black people only	3	6.8%
All of the above*	358	89.5%
Total	400	100%
Effects of SCD on the red blood cell		
Round and soft	38	9.6%
Hard and sickle shaped	111	27.8%
Sticky and blue	35	8.2%
Stiff and sickle shaped*	106	26.2%
Soft and sickle shaped	112	28.2%
Total	400	100%

Table 4.2 continued		
variables	N	%
Possible management of SCD		
Use of antibiotics	61	15.2%
Liver transplant	63	15.9%
Use of pain relievers	137	34.2%
Blood transfusion	53	13.2%
None of the above	86	21.5%
Total	400	100%

	N	%
Cure of SCD		
Bone marrow transplants*	114	28.5%
Antibiotics	83	28.8%
Rest	53	13.2%
All of the above	145	36.2%
None of the above	5	1.2%
Total	400	100%
Possible diagnosis of SCD		
Sickling appearance	126	31.5%
Simple blood test	225	63.8%
Impossible to diagnose	12	3.0%
Patients diseased look	7	1.8%
Total	400	100%
Prevention of SCD		
Use of condom	10	2.50%
Premarital genetic testing and counselling*	314	78.5%
Abortion	11	2.8%
All of the above	14	3.5%
None of the above	51	12.8%
Total	400	100%

Appropriate testing time	N	%
When feeling sick	60	15.0%
At any time	172	43.0%
When one is ready to wed	92	23.0%
All of the above	76	19.0%
Total	400	100%
Purpose of premarital testing		
Knowledge of sickle cell trait status	113	28.2%
Prevention of incidence of Sickle cell disorder	96	24.0%
To make an informed marital choice	115	28.8%
None of the above	4	1.0%
All of the above	72	18.0%
Total	400	100%

Table 4.3 Knowledge Score

Grade	Value	No	%	Mean ±SD
Poor	≤ 10	64	16.0	
fair	>10-20	252	63.0	11.4 ± 5.0
Good	>20	84	21.0	
Total		400	100.0	

4.3 Utilisation of Premarital Sickle Cell Genetic Testing and Counselling Services

This section presents utilization of Sickle Cell Counselling and Testing. Majority (97.0%) of the respondents knew that one can check his sickle cell status while only 3.0% had no knowledge that one can check his status. Ninety-seven percent of the respondents were aware of the availability of the sickle cell testing service (97.0%), while (3.0%) did not know about the availability of the service. Meanwhile, 5.0% mentioned family guidance and counselling centres, 10.0% mentioned private clinics, while most (85.0%) of the respondents mentioned government hospitals.

Equal proportion (27.0%) of the respondents live between 2km to 3km and more than 3km away from service centres, while 33.0% live between 1km – 2km away from service centre and 13.0% live less than 1km from service centre (table 4.4)

Majority (61.5%) of the respondents had done Sickle Cell testing, while 38.5% did not consider it an option. Of the respondents that had done the testing, 10.0% of the respondents tested because of significant others; a similar proportion did testing because of statutory marriage pre-requisite from their church, while 30.0% of the respondents opted for testing because of the possible consequences of marriage between two carriers. However, 50.0% of the respondents considered all the mentioned options.

Out of the 246 respondents that have done testing (61.5%), (2.0%) did the test at youth friendly centre, majority had used the general hospital (60.0%), (3.0%) used the church, (22.0%) used the college clinic, while (13.0%) did the test at the laboratory (Table 4.4)

Majority 195 (79.0%) of the participants that have done the test paid for the test. while less than half 51(21.0%) of the respondents did the test for free. Fifty-eight percent of the participants paid between N300 - N500 for the test (58.0%), five percent paid between less than N100 to N200 (5.0%) and majority of the participants paid between N300 to N500, while 24.0% paid more than N500 to do the test. A substantial proportion (84.0%) was of the view that SCT is affordable, while a minority felt that it was expensive. .(44.5%) said the test was not time consuming, (52.8%) said it was time consuming, while only (2.8%) said it was too time consuming.

Sixty percent of respondents who had done testing were asked about the attitude of the health care workers that attended to them, 60.2% reported that healthcare workers were friendly, while 39.8% commented that they were unfriendly.

Table 4.4 Respondents' Utilisation-Related Statements to SCT.

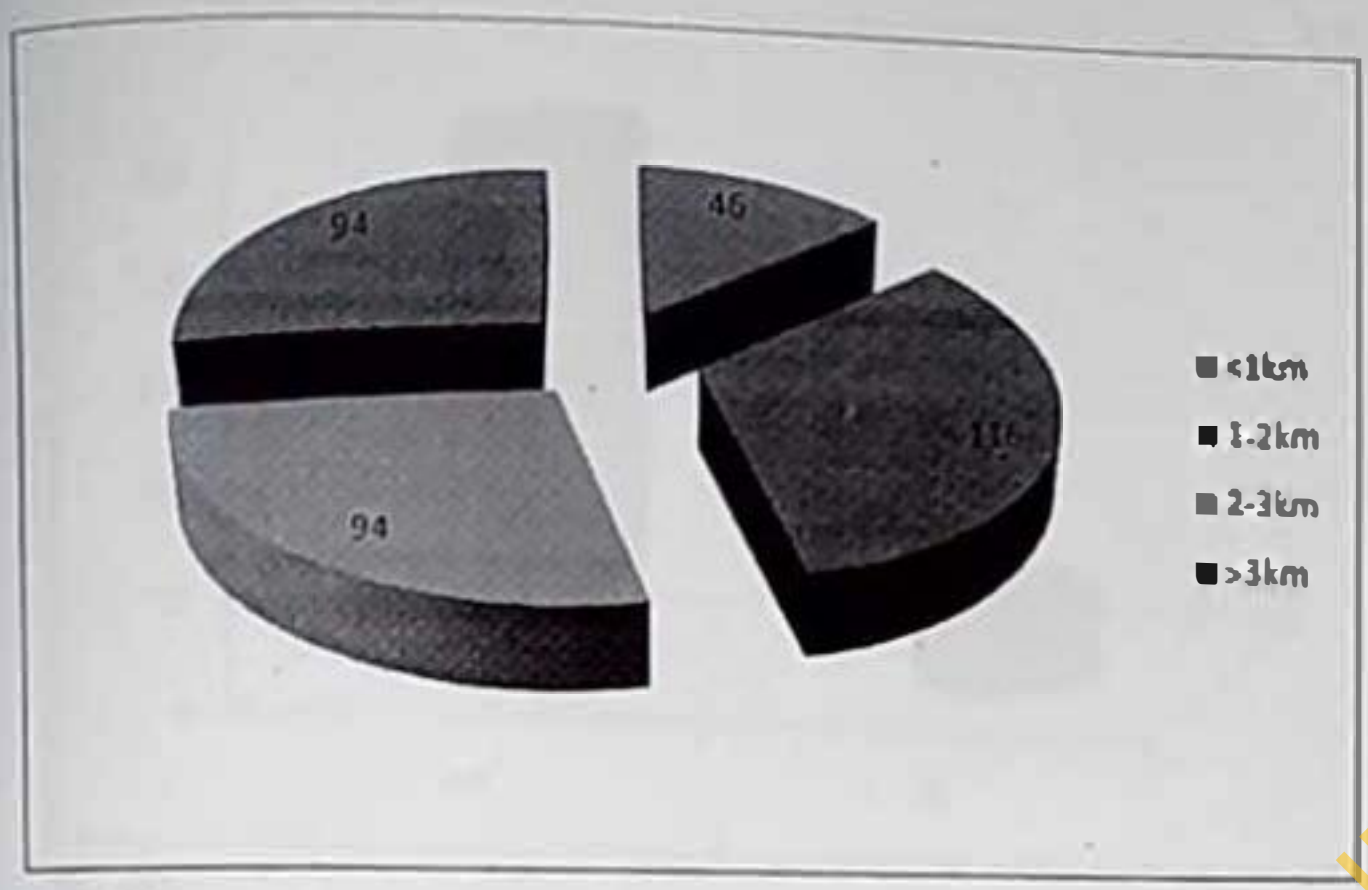
Variables	N	%
Do you know that one can check his SC status (N=400)		
Yes	388	97.0%
No	12	3.0%
Total	400	100%
Do you know SC testing sites (N=388)		
Yes	350	90.5%
No	38	9.5%
Total	388	100%
Sites where test could be done (N= 350)		
Family guidance and counselling centre	17	5.0%
Private clinics	35	10.0%
Government hospitals	298	85.0%
Total	350	100%
Shortest distance of testing site (N=350)		
< 1km	46	13.0%
1 – 2km	116	33.0%
2 – 3km	94	27.0%
>3km	94	27.0%
Total	350	100%
Have you ever done any form of genetic testing N= (400)		
Yes	246	61.5%
No	154	38.5%
Total	400	100%

Table 4.4 contd

Variables	N	%
What made you consider testing (N=246)		
Significant others	24	10%
Marriage pre-requisite	24	10%
Possible consequences of SCD	75	30%
All of the above	123	50%
Total	246	100%
Where did you do the testing? (N=246)		
Youth friendly centre	5	2.0%
General hospital	148	60.0%
Church	7	3.0%
University clinic	49	22.0%
Private laboratory	32	13.0%
Total	246	100%
Was the test free of charge		
Yes	12	6.0%
No	232	94.0%
Total	246	100%
How much did you pay for the test (N= 194)		
<N100	10	5.0%
N100-N200	10	5.0%
N200-N300	16	8.0%
N300-N500	112	58.0%
>N500	46	24.0%
Total	194	100%
Would you consider the test cost affordable?(N=194)		
Yes	163	84.0%
No	31	16.0%
Total	194	100%

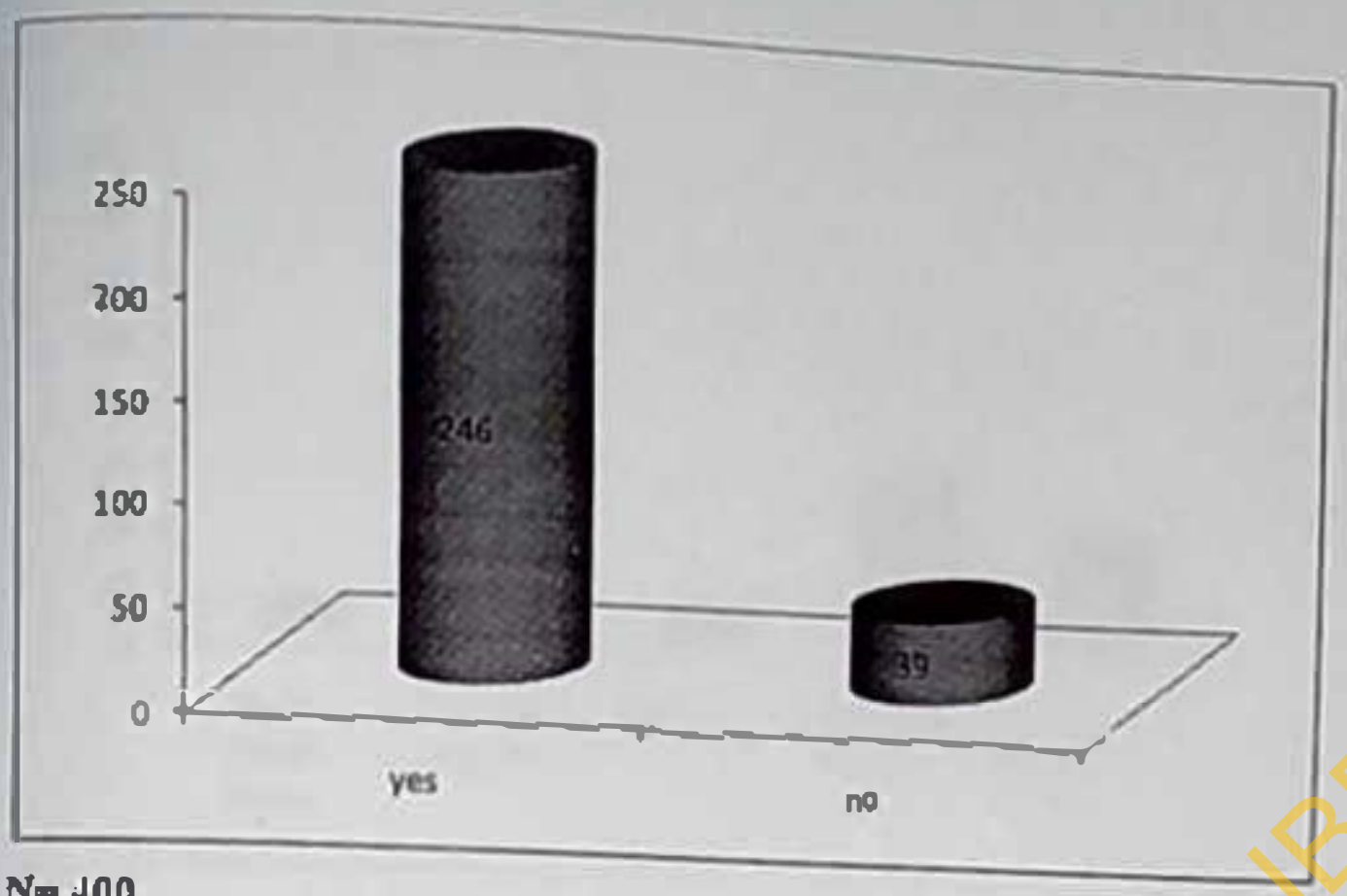
Duration of the test (246)		
Not time consuming	109	44.5%
Time consuming	130	52.8%
Too time consuming	7	2.8%
Total	246	100%
Health care workers attitude (246)		
Friendly	148	60.2%
Unfriendly	98	39.8%
Total	246	100%

Duration of the test (246)		
Not time consuming	109	44.5%
Time consuming	130	52.8%
Too time consuming	7	2.8%
Total	246	100%
Health care workers attitude (246)		
Friendly	148	60.2%
Unfriendly	98	39.8%
Total	246	100%



N= 350

Fig 4.2 Participants' Response Relating to Distance of Service Centres



N= 400

Fig 4.3 Prevalence of Utilization

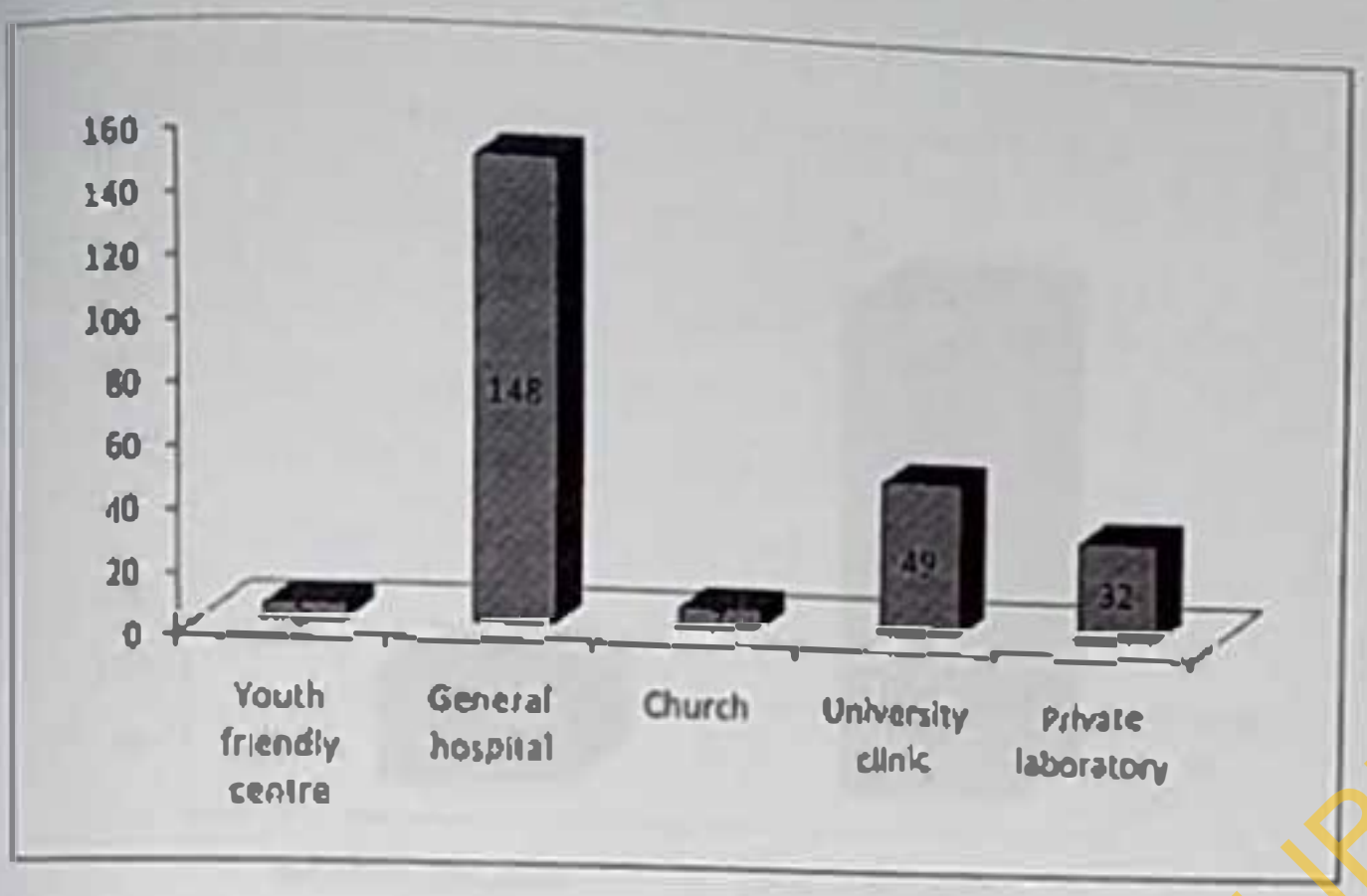


Fig 4. 4 **Distribution of Service Centres** N = 246

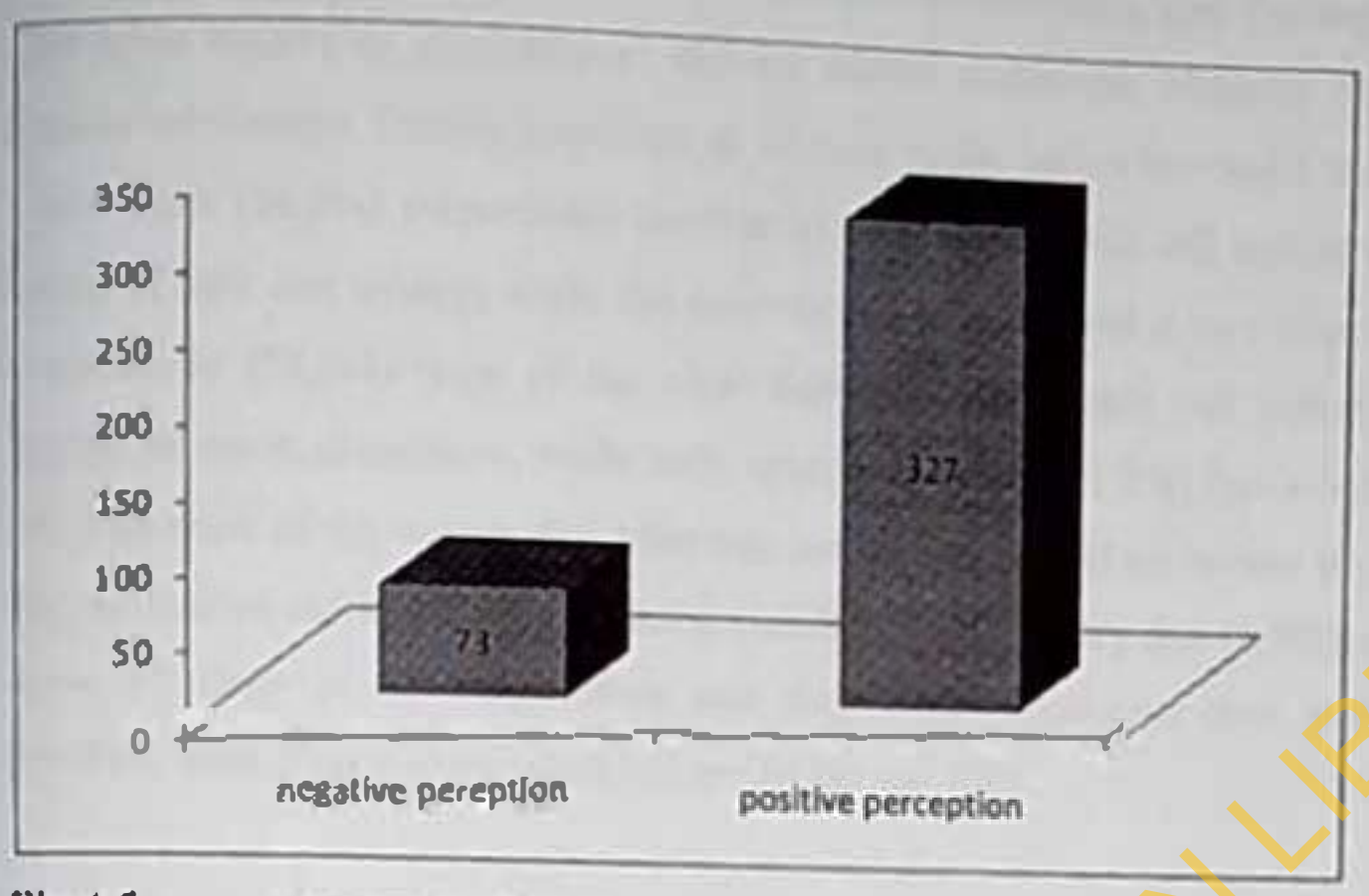


Fig 4.5 Perception Score

4.4 Attitude towards Premarital Sickle Cell Counselling and Testing

The table highlights respondents' attitude related statement. Majority (93.2%) considered Premarital Genetic Testing important at all cost, while only a few had a contrary perception. Only a few (18.0%) respondents considered premarital sickle cell testing and counselling a waste of time and money, while the majority (82.0%) deemed it very important. Most of the respondents (78.5%) were of the view that premarital sickle cell testing and counselling should be made mandatory, while only small proportion (21.5%) had a contrary view. Most (91.5%) were of the viewer that Marriage institutions should encourage their members to go for pre-marital sickle cell testing and counselling, while a very few (8.5%) were of a contrary view. Majority of the respondents said they would encourage their partner to be tested (90.0%), even if they were tested to have sickle cell trait.

Attitude scale was used to assess the attitude of the respondents towards PSCCT. Respondent who scored 10, were rated as having negative attitude and respondents with >10 were rated as having good knowledge. Majority of the respondents (89.0%) had good attitude towards PSCCT compared to those who were rated to have negative attitude. In conclusion, the mean attitude score of respondents was 7.4 ± 1.8 (Table 4.5 and Figure 4.6).

Majority of the IDI discussants agreed that PSCCT should be mandatory rather than optional. Their typical responses include the following.

- *If a country like Cyprus tried mandatory of Premarital screening and it reduced incidence of genetic disorder, particularly the Sickle cell disorder and some part of South is adopting the some despite the possibility of consanguinity marriage. I see no reason why country like Nigeria should not welcome it with a good applause*
- *Honestly, PSCCT should not be seen as an accessory for marriage, but prerequisite for marriage.*
- *Proper orientation should be given to intending couples with respect to PSCCT so that they can appreciate its benefit after marriage*

Table 4.5 Respondents Attitude towards PSCCT (N=400)

Variables	N	%
Do you consider premarital sickle cell testing important at all cost		
Yes	372	93.2%
No	28	6.7%
Total	400	100%
Premarital Sickle Cell Testing and Counselling is a Waste of Time		
Yes	72	18.0%
No	328	82.0%
Total	400	100%
Premarital Sickle Cell Testing should be made mandatory		
Yes	314	78.5%
No	86	21.5%
Total	400	100%
Marriage institutions should encourage their members to go for pre-marital sickle cell testing and counselling		
Yes	366	91.5%
No	34	8.5%
Total	400	100%
Would you encourage your spouse to go for premarital sickle cell testing		
Yes	360	90.%
No	40	10.%
Total	400	100%

Table 4.6 Attitude Score

Grade	Value	No	%	Mean ±SD
Negative attitude	≤ 10	44	11.0	
Positive attitude	>10	356	89.0	7.1±1.8
Total		400	100.0	

4.5 Respondents' Perception of Sickle Cell Disorder and Health Beliefs

Majority (89.8%) of the respondents agreed that Sickle Cell Disease is a serious disease, while a negligible (2.8%) proportion disagreed. A substantial proportion (70.5%) of the respondents strongly agreed that having a Child with SCD would be very scary, while only a small proportion (13.5%) disagreed strongly. Above half 77.0% of the respondents had the perception that Children with sickle cell disease are at risk for infections, Pain, pneumonia, while the minority (8.2%), strongly disagreed. Most (85.5%) of the respondents were of the view that if both partners are carriers of sickle cell trait, children could be at risk. Sickle Cell Trait is transmittable to offspring. Slightly above half (60.0%) of the respondents had the perception that anyone may be a carrier of sickle cell trait, while only 24.8% strongly disagreed with such perception.

A large (84.2%) proportion of the respondents agreed that Sickle Cell Disorder can be prevented in offspring if both partners know their SCT status, but only 7.0% were of a contrary view. Most (84.0%) of the respondents had the perception that knowledge of the burden of having a child with SCD would make them engage in pre-marital SCT, while only (12.8%) strongly disagreed with such perception. A few (13.2%) respondents were of the view that Testing for sickle cell trait is painful and difficult, while some (63.3%) of them disagreed with such perception. Only (12.8%) said they would not want to pay for SCT, because they considered it expensive, while slightly above half of the respondents did not consider it expensive.

Only a small (18.8%) proportion were worried that their screening result could get into outsider's hands, while about half (55.5%) of the respondents were not anxious about result getting into outsiders' hnnd. About half (55.8%) of the respondents did not feel that genetic testing could lead to discrimination hence, did not accept testing, while only 21.0% strongly felt testing could lead to discrimination. A quarter (25.0%) of the respondents were of the opinion that service centres were distant to their residence, while half (50.0%) did not agree that service centres were distant to their residence. Minority (13.0%) strongly agreed that their culture does not support premarital sickle cell testing and counselling. while a larger (76.0%) proportion were of a contrary opinion. Similarly, only a small (13.2%) proportion said genetic testing contradicts their faith.

Perception scale was used to assess the perception of perception about PSCCT. Respondents, who scored 10, were rated as having poor knowledge. Those who had > 10 were rated as having positive pe4rception. Majority of the respondents (81.8%) had positive perception in

compared to (18.2%) of the respondents who had negative perception. In conclusion, the mean perception score of respondents was 12.4 ± 3.8. (Table 4.7 and Figure 4.6).

Table 4.7 Respondents Perception of Sickle Cell Disorder and Health Beliefs (N=400)

Variables	N	%
Sickle Cell Disease is a serious disease		
Strongly agree	356	89.8%
Undecided	30	7.5%
Strongly disagree	11	2.8%
Total	400	100
Having a Child with SCD would be very scary		
Strongly agree	282	70.5%
Undecided	64	16.0%
Strongly disagree	54	13.5%
Total	400	100%
My life would change if my child had SCD		
Strongly agree	205	51.2%
Undecided	113	28.3%
Strongly disagree	82	20.5%
Total	400	100%
Children with sickle cell disease are at risk for Infections, Pain, pneumonia		
Strongly agree	308	77.0%
Undecided	56	14.0%
Strongly disagree	36	8.2%
Total	400	100%
If both parents are carriers of sickle cell trait, children could be at risk		
Strongly agree	342	85.5%
Undecided	28	7.0%
Strongly disagree	30	7.5%
Total	400	100%
Sickle Cell Trait is transmittable to offspring		
Strongly agree	296	73.8%
Undecided	44	11.0%
Strongly disagree	60	15.0%
Total	400	100%

Variables	N	%
Anyone may be a carrier of sickle cell trait		
Strongly agree	240	60.0%
Undecided	61	15.2%
Strongly disagree	99	24.8%
Total	400	100%
SCD can be prevented if both partners know their status		
Strongly agree	337	84.2%
Undecided	35	8.8%
Strongly disagree	28	7.0%
Total	400	100%
Knowing my sickle cell trait will make me confident in forming relationship		
Strongly agree	336	84.0%
Undecided	29	7.2%%
Strongly disagree	35	8.8%
Total	400	100%
Knowing the burden of having a child with SCD would make me go for PSCCT		
Strongly agree	301	75.2%
Undecided	48	12.0%
Strongly disagree	51	12.8%
Total	400	100%
Testing for SCT is painful and difficult		
Strongly agree	53	13.2%
Undecided	94	23.5%
Strongly disagree	253	63.3%
Total	400	100%
It would be difficult convincing my wife to have testing		
Strongly agree	88	22.0%
Undecided	100	25.0%
Strongly disagree	212	53.0%
Total	400	100%

Table 4.7 contd Respondents perception of sickle cell disorder and health beliefs(N=400)

Variables	N	%
I would not want to pay for PSCCT because it is expensive		
Strongly agree	51	12.8%
Undecided	97	24.2%
Strongly disagree	252	63.0%
Total	400	100%
I am worried that the test results could get into outsider's hand		
Strongly agree	75	18.8%
Undecided	103	25.8%
Strongly disagree	222	55.5%
Total	400	100%
Genetic testing is not acceptable because the result could lead to discrimination		
Strongly agree	84	21.0%
Undecided	93	23.2%
Strongly disagree	223	55.8%
Total	400	100%
Service centre are distant to my residence		
Strongly agree	100	25.0%
Undecided	100	25.0%
Strongly disagree	200	50.0%
Total	400	100%
My culture does not support PSCCT		
Strongly agree	52	13.0%
Undecided	44	11.0%
Strongly disagree	304	76.0%
Total	400	100%
Genetic testing is not acceptable by my faith	53	13.20%
Strongly agree	48	12.0%
Undecided	299	74.8%
Strongly disagree	400	100%
Total		

Table 4.7 contd Respondents perception of sickle cell disorder and health beliefs(N=400)

Variables	N	%
I would not want to pay for PSCCT because it is expensive		
Strongly agree	51	12.8%
Undecided	97	24.2%
Strongly disagree	252	63.0%
Total	400	100%
I am worried that the test results could get into outsider's hand		
Strongly agree	75	18.8%
Undecided	103	25.8%
Strongly disagree	222	55.5%
Total	400	100%
Genetic testing is not acceptable because the result could lead to discrimination		
Strongly agree	84	21.0%
Undecided	93	23.2%
Strongly disagree	223	55.8%
Total	400	100%
Service centre are distant to my residence		
Strongly agree	100	25.0%
Undecided	100	25.0%
Strongly disagree	200	50.0%
Total	400	100%
My culture does not support PSCCT		
Strongly agree	52	13.0%
Undecided	44	11.0%
Strongly disagree	304	76.0%
Total	400	100%
Genetic testing is not acceptable by my faith	53	13.20%
Strongly agree	48	12.0%
Undecided	299	74.8%
Strongly disagree	400	100%
Total		

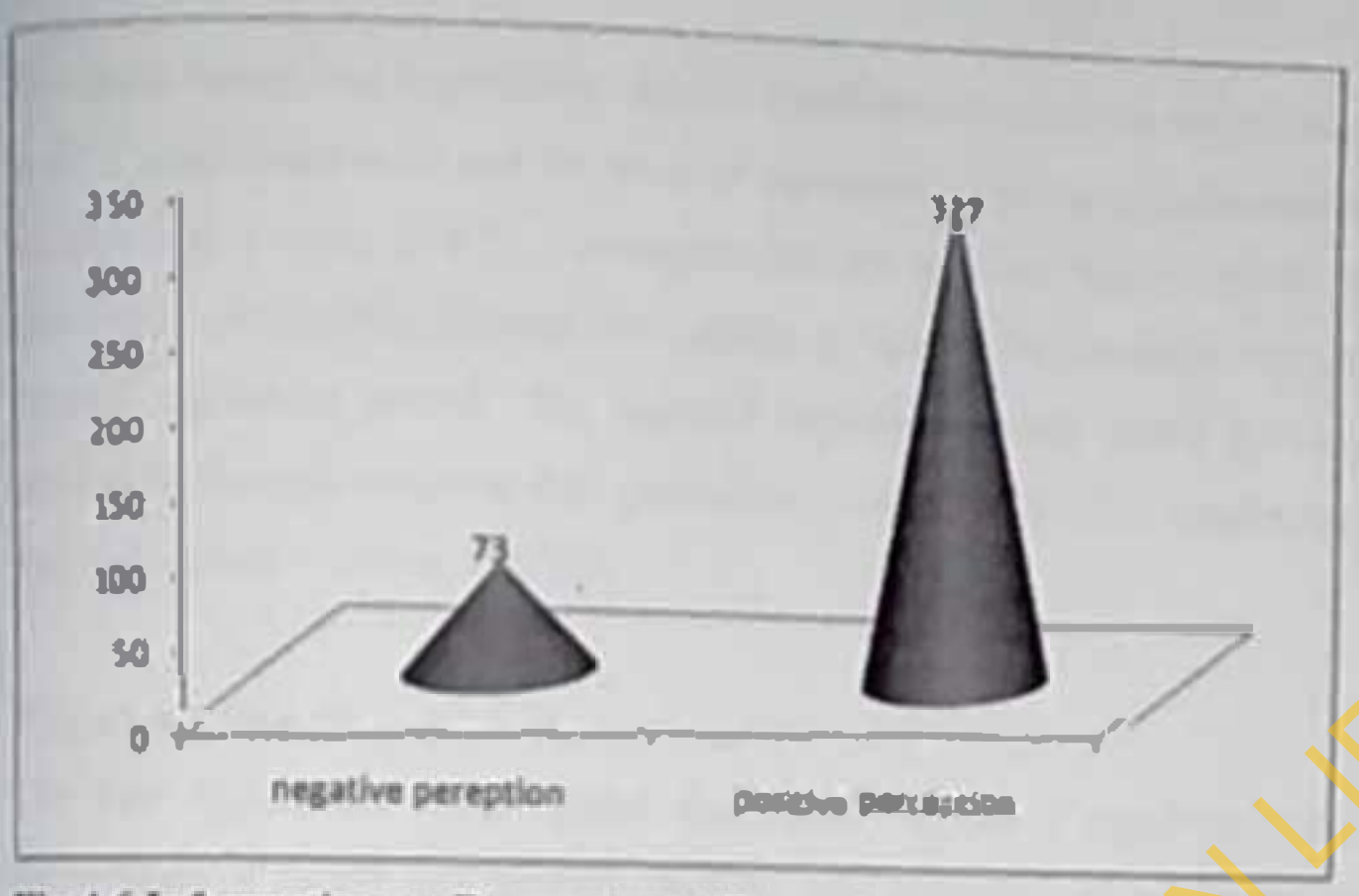


Fig 4.6 Information on Respondents' Perception

4.6 Test of Hypotheses

The study tested four hypotheses. All the hypotheses were proposed as alternative hypotheses at 95% confidence level and 5% level of statistical significance otherwise referred to as limit of error with p value of 0.05. The appropriate test statistics used to determine the existence or absence of relationship between the variable of interest for the study was Chi-square (X^2) and logistic regression model. The research hypotheses were tested by comparison between utilisation through variables like, perception, level of education, knowledge of SCD, attitude and willingness to utilise PSCCT.

Hypothesis one (H_1)

The first hypothesis which stated that, there will be a significant relationship between knowledge of sickle cell genetic disorder and utilisation of premarital sickle cell genetic counselling and testing services.

Table 4.8 shows the cross tabulation of respondents' level knowledge of SCD with utilisation using Chi-Square test. There was a significant relationship between respondent level of knowledge and utilization of PSCCT at 95percent confidence interval ($p<0.05$). Respondents' knowledge has a role to play in the utilisation of PSCCT. The null hypothesis was therefore rejected.

Hypothesis two (H_2)

The second alternative hypothesis which stated that there will be a significant relationship between perception and utilization of premarital sickle cell genetic counselling and testing. Table 4.9 shows the cross tabulation of perception and utilisation of PSCCT using Chi-Square statistic. There was a significant relationship between perception and utilisation of PSCCT at 95 per cent confidence interval ($p<0.05$). Respondents' perception has a role to play in utilisation of PSCCT. The null hypothesis was therefore not rejected.

Table 4.9 shows the cross tabulation of respondents' highest level of education with respondents' utilization of PSCCT using Chi-Square test. There was a significant relationship between respondents' highest level of education and prevalence of utilization at 95 per cent

confidence interval ($p<0.05$). Respondents' level of education is a key factor in determining utilization of PSCCT. The null hypothesis was therefore rejected.

Hypothesis three (H_3)

The third alternative hypothesis which stated that there will be a relationship between attitude and utilisation of premarital sickle cell genetic counselling and testing services Table 4.10 shows the cross tabulation of attitude and utilisation of PSCCT using Chi-Square statistic. There was a significant relationship between attitude and utilisation of PSCCT at 95 per cent confidence interval ($p<0.05$). Respondents' attitude has a role to play in utilisation of PSCCT. The null hypothesis was therefore rejected.

Hypothesis four (H_4)

The fourth alternative hypothesis which states that, there will be a significant relationship between level of education and utilisation of premarital sickle cell counselling and testing. Table 4.11 shows the cross tabulation of attitude and utilisation of PSCCT using Chi-Square statistic. There was a significant relationship between attitude and utilisation of PSCCT at 95 per cent confidence interval ($p<0.05$). Respondents' level of education has a pivotal role to play in utilisation of PSCCT. The null hypothesis was therefore rejected.

Table 4.8 Level of knowledge of SCD and Utilisation PSCCT

Knowledge score	utilisation of PSCCT		
	Strongly agree Freq. (%)	Undecided Freq. (%)	Strongly disagree Freq. (%)
Poor	38 (9.5)	15(3.8)	11(2.8)
Fair	190(47.5)	28(7.0)	34 (8.5)
Good	73(18.3)	5(0.8)	6 (33.5)
Total	301(75.3)	48(12)	51 (12.75)

$\chi^2 = 16.532$

df = 4

p = 0.002

Table 4.9 Perception and Utilisation of PSCCT

Perception score	Utilisation of PSCCT		
	Poor utilisation Freq. (%)	Fair utilisation Freq. (%)	Good utilisation Freq. (%)
Poor	2(0.5)	32(8)	39(9.8)
Good	0(0)	5(0.8)	6 (33.5)
Total	2(0.5)	37(9.3)	45 (11.2)

$\chi^2 = 10.706$

df = 2

p = 0.004

Table 4.10 Relationship between Attitude and Utilisation of PSCCT

Attitude score	Utilization score		
	Poor utilisation Freq. (%)	Fair utilisation Freq. (%)	Good utilisation Freq. (%)
Negative attitude	2(0.5)	26(6.5)	16(4.0)
Positive attitude	0(0.0)	126(31.5)	230(57.5)
Total	2(0.5)	152(38.0)	246(24.3)

$\chi^2 = 27.048$

df = 2

p = 0.000

Table 4.11 Level of Education and Prevalence of Utilisation of PSCCT

Level of education	Have ever done sickle cell genetic testing	
	Yes Freq. (%)	No Freq. (%)
Primary	4(1.0)	5(1.3)
Secondary	37(9.3)	30(7.5)
Tertiary	201(50.3)	115(28.8)
No formal	4(1.0)	4(1.0)
Total	246 (61.5)	154 (38.5)

$\chi^2 = 18.693$

df = 4

p = 0.001

4.12 Binary Logistic Regression

Categorically, after effecting adjustment, it was revealed that respondents with fair knowledge of SCD were more 2.5 more likely to utilise PSCCT compared to those with poor knowledge (Table 4.12).

Table 4.12 Binary Logistic Regression on knowledge about SCD

Variable	Odds ratio (95% CI)	P-value
Knowledge score		
Fair knowledge score (Reference)	2.5	p = 0.001
Poor knowledge	(2.3 – 4.2)	

CHAPTER FIVE

DISCUSSION, CONCLUSION AND RECOMMENDATIONS

Knowledge of the citizenry of a nation about SCD constitutes an important variable that influences the acceptability, practice and success of premarital genetic counselling. Many young women and men enter into marriage with insufficient information on sexuality, reproduction, and family planning (*Bastani etal; 2010*). This study focused mainly on about to wed or about-to-wed couples because their beliefs and attitudes will affect their choices in life.

5.1 Socio-Demographic Characteristics

The age of respondents ranges from 20 to 30 and above, which is similar to age range used by (*Abioye etal; 2009*) in a related studies among Local Government workers in Ile Ife. The selected respondents made it possible to investigate knowledge, perception and attitude of the about-to-wed couples towards premarital sickle cell genetic testing and counselling. In terms of gender difference, 45.0% were male and 55.0% female showing that the participation was higher with female respondents. This is in line with *Durojaye etal; 2013* where there (45.6%) male and (55.5%) female. This shows that a reasonable percentage were self-employed which is not unconnected with the unemployment rate (23.9%) in the country as documented by National Bureau of Statistics 2011.

Concerning level of education, (2.0%) had no formal education (2.2%) attained primary education, (6.6%) attained secondary education while (79.0%) attained tertiary education. This was in line to the outcome of a related study among local government workers in Ile – Ife by (*Abioye etal; 2009*) where the majority (65.7%) of the respondents had tertiary education. With regards to religion majority (47.0%) were Moslems and (52.0%) were Christians. This is probably due to the fact that the study location-Ibadan has a fairly high percentage of Christian faithful than Muslim.

5.2 Level of Knowledge SCD among Respondents

The IDI findings showed that both male and female respondents have generally fair knowledge about SCD; male and female alike. The population of female during the IDI sessions outnumbered the male respondents and interactions showed that majority, if not all were educated.

The relationship between respondents' knowledge about sickle cell disorder and the demographic characteristics concerning age; shows that respondents between ages 20 – 25 had highest good knowledge score (26.9%), while respondents above 30 years of age had highest proportion of fair knowledge score (68.8%).

With regards to level of education, respondents with formal education had highest fair knowledge score, unlike respondents with no formal education. This shows that education is instuumental to better knowledge about SCD. This result is at variant with a study Durotoye etal; 2013 findings, which was conducted among SS3 students. The difference could be ascribed to the maturity discrepancies between the respondents of the two studies in which younger students lack the understanding of seriousness of the genetic blood disorders and its huge impact on the emotional and financial status of the affected families.

Based on ethnicity, Yoruba respondents had highest (65.7%) percentage of fair knowledge score compared to other ethnic group with the highest (34.5%) good knowledge score. In terms of religion, both the Christians and the Muslims had a similar fair and good knowledge score. In terms of the living status, respondents living with alone had the highest percentage (21.1%) of good knowledge score and followed by respondents living with their parents (20.1%).

Differences from the studies were ascribed to difference in education level of the study participants. A good proportion (76.0%) knew the cause of SCD as an hereditary disease, which is not in line with *Durotoye etal; 2013* findings, which was conducted among SS3 students. The difference could be related to the maturity discrepancies between the respondents of the two studies where they lack the understanding of seriousness of the genetic blood disorders and its huge impact on the emotional and financial status of the affected families.

From this study, Respondents that had tertiary (79.0%) education had the highest proportion of (84.8%) favourable knowledge score of which is at variant with *Abioye etal; 2012* findings in which only a small proportion (31.0%) of respondents had good knowledge of SCD, despite being the majority (69.7%).

Based on gender, women also had better knowledge (56.7%) and stronger attitudes toward the implementation of screening (73.6%). This is in support of a study by *Arulogun and Adefioye*; 2010, found out that woman tend to have more knowledge (61.3%) about issues relating to reproductive and sexual health than their male counterparts.

Despite the good knowledge of SCD by the respondents of this study, major proportion (71.5%) did not know that bone marrow transplantation is the only known curative measure for SCD. This result is consistent with (*Durotoye etal; 2013*) and a similar study in Benin in which (61.0%) of the respondents believed that SCD is incurable (*Baruwa etal, 2009*).

In Nigeria, there is a facility for stem cell transplantation in Benin city, Edo state and to date, three sicklers have undergone the procedure and currently doing well. The major challenge of this treatment is the high cost of the procedure and difficulty in finding suitable donors.

5.3 Attitude of Respondents towards Premarital Sickle Cell Counselling and Testing.

IDI discussions showed that female folks had better attitude towards utilisation of PSCCT saying that, it is very important to know ones state of health. Most of the women that had not done testing promised to conclude all their premarital testing as soon as possible.

From this study, a favourable attitude score of 89.0% was documented. However, female respondents had greater proportion (90.5%) of favourable attitude score, this is in consonance with a similar study of *(Al-Aama etal; 2011)* on attitudes toward mandatory national premarital screening for hereditary haemolytic disorders discovered that women also had better knowledge (56.7%) and stronger attitudes toward the implementation of screening (73.6%). Also, *Arulogun and Adefioye*; 2010, found out that women tend to have more knowledge (61.3%) about issues relating to reproductive and sexual health than their male counterparts as a result, women folks tend to have positive attitude towards their sexual health. Similar studies such as those by *Khater and El Ghazaly; 2003 and Abd Al-Azeemet al; 2012* demonstrated that women were more oriented and as such more knowledgeable (67.4 and 62.3%, respectively) with important health issues related to premarital genetic screening than men (53% and 47%, respectively), which reflected on their better attitude.

Concerning attitude towards premarital screening, majority (78.5%) agreed that premarital sickle cell screening should be made mandatory. The same result was obtained from a similar study among University students, majority of the participants' attitudes towards PMS were favourable, where the majority believed that PMS is important and agreed to carry it out in the future. This is also similar to what has been reported in other Arab countries (*Eshraet al. 1989; Al-Aama etal, 2008, Al-Kahtani etal, 2009*).The majority of the participants reported that they will perform Premarital Screening(PMS) to prevent transmission of diseases to their children. This reflects that the participants had a good understanding of the preventive value of PMS *(Al Kindi etal, 2012)*

Regarding ethnicity, Yoruba respondents were the largest proportion (89.4%) in terms of good attitude score. Educationally, respondents with tertiary education were the highest proportion (90.2%) with good attitude score. This is no doubt a pointer to the fact that knowledge has a direct relationship with attitude towards premarital sickle cell testing. Some people believe that their fate is determined by God and therefore accept the risk of having a sick child.

5.4 Perception of Sickle Cell Disorder

The perception of the female IDI discussants was very high, because most of them said that, the women are always the pillar of the house when it comes to children's health. They corroborated that, men do not have time. Therefore, women will always bear the brunt if anything happens to the children in future.

This study documented a favourable perception percentage score of 51.8%, which had a higher proportion (83.7%) of female respondents. It shows that gender plays an important role in the perception. This result is consistent with findings of *Al-Aamo (2010)* in a study on attitudes towards mandatory national premarital screening for hereditary haemolytic disorders discovers that women also had better disposition towards the implementation of screening with a significantly higher number of female respondents believing that the pre-marital screening should be mandatory and that marriage should not be allowed between two carriers of the same disorder. They speculated that the discovery may reflect the belief that women in some societies may bear more of the burden of a handicapped or chronically ill child and they have less opportunity to a second chance than men.

Similarly, Arulogun and Adefioye *(2010)* find out that females tend to have more knowledge about issues relating to reproductive and sexual health than their male counterparts. The reason they adduced to this is the fact that many of the Christian religious organizations have a compulsory counselling sessions for intending couples which last for months before the marriage is conducted. In these counselling sessions, the intending couples are educated on issues regarding marriage and the home; whereas, in most religious places in Nigeria, especially in churches, there are usually more women than men.

In terms of living status, respondents living with other relatives had highest proportion of good perception. This could result from influence of possible significant others as regards Premarital screening decisions.

5.5 Utilisation of Premarital Sickle Cell Genetic Counselling and Testing

The study recorded a favourable utilization score of 61.5%.Female respondents had higher proportion of utilization compared to men. There is a direct relationship between perception and attitude which in turn increases along the trend of utilization. Age wise, respondents between the age of 26 – 30years had the highest utilization percentage compared to respondents above 30years. On the contrary, findings of a related study showed that older individuals appear to have higher rates of participation in premarital screening (*Peigang etal; 2013*). This may be due to the fact that, as the age increases, people attach increased importance to their health. An increasing sense of responsibility with age may also influence participation (*Misri and Munla, 2004*). Increased concern about birth defects may also be partially responsible.

Professionally, the cultural literacy and professional characteristics of the scientists and teachers, may give them a more comprehensive and clearer understanding of PMS.

5.6 Implications to Health Promotion and Education

Educational programs need to be constantly upgraded by literature and research findings as regards to improving establishment of genetic counselling and testing centres and also special unit for genetic counselling and testing apart from the conventional haematology department. Health education should include the latest recommended evidence based literature most especially in area of statistics on the utilization of genetic screening. The evidence from the present study indicated a good knowledge of SCD, but most of the screening facilities are restricted to urban areas. There is a need at all level to make facilities for genotype screening and genetic counselling evenly distributed at both urban and rural communities where many people reside.

There is the need for policy in the Country preventing carriers from marrying each other, and the need for constant education for the youth to take useful decision l mate choices as financial implication of managing a sickler which is a long life ailment can cause disharmony in the family.

Ultimately, there should be a policy support by Government to promote in all marriage to promote Premarital Sickle Cell Counselling and Testing units in all marriage institutions: religious, traditional and courts.

5.7 Conclusion

The findings from this study showed that majority of the respondents were within the age range of 26 – 30 years, majority attained tertiary education level, and were in paid employment. Most of them were Christians and the majority of the respondents were Yoruba. The respondents had a good knowledge of Premarital Sickle Cell Genetic Disorder. The attitude, perception and utilization of the service by the respondents were good.

The study is limited to only about-to-wed about to wed couples who contracted their wedding at Ibadan North East Local Government Registry.

5.8 Recommendations

1. Implementation of school and university educational campaigns is important. Improved counselling and adding new topics for counselling on genetic and chronic problems; building healthy families; reproduction and fertility are recommended.

2. If sickle cell disease control strategies must yield any significant results, more education about SCD, especially among secondary school students in Nigeria is therefore recommended. The use of persons with SCD as peer educators/counsellors should be explored.

3. Involvement of community leaders and non-governmental organizations in counselling programs to youth to raise awareness and change their attitude toward marriage between genetically incompatible partners.

4. Religious organizations can serve as useful channel to disseminate this health information since people always have reference for places of worship and for religious leaders.

5. Ethical principles of justice, autonomy, confidentiality, beneficence and respect for the dignity and basic intelligence of persons should be adhered to because this will go a long way in helping people to embrace pre-marital genetic screening without any reservation.

6. The health beliefs, traditions, religious observances and social expectations of individuals and communities should be assessed properly before setting program

goals, and respected thereafter. These goals should never be set in ways to impose certain genetic screening or reproductive decisions on individuals.

7. To encourage a transformation from passive knowledge in pre-marital genetic screening gained through accidental learning, there is an urgent need for the inclusion of Health Education in the curriculum of Nigerian tertiary education to be taught as a course regardless of discipline.

8. Governments should recognize that within any country there exists diversity of cultures and opinions about n number of issues relevant to genetics. These include human reproductive issues, community and individual approaches to the significance of disabilities, this diversity should be respected.

9. The meaning of the term 'carrier status' should be made known to members of the public long before they get married. For successful public education, government and noo-governmental organizations must cooperate, as well as community and religious leaders, school parent organizations and health personnel.

10. At-risk couples should be contacted directly by counsellors and encouraged to undertake education and counselling.

11. Although, prevention and treatment of genetic diseases is virtually impossible. There is the need for physicians to offer counselling for various health problems before the patient asks, so they can turn down unwanted help rather bring up sensitive issues.

12. Implementation of 'solution-focused' premarital counselling that focuses on a couple's resources, helping them to develop a shared vision for the marriage. Background information about premarital counselling and solution-focused therapy to provide a framework in which intervention strategies in those confirmed with positive status for a disease can be developed

13. A solution-oriented interventions include solution-oriented questions and feedback, as well as a Couple's Resource Map (CRM) which depicts the support available to the couple from various personal, relationship and contextual resources

14. Screening programmes must be equitable, accessible and understood by the target population, but most importantly they must comply with the prevailing cultural, ethnic, economic and societal values.

REFERENCES

Abioye-Kuteyi E. A, Oyegbade O, Bello I,Osakwe C. Sickle cell knowledge, premarital screening and marital decisions among local government workers in Ile-Ife, Nigeria. Afr J Prm Health Care Fam Med. 2009;1(1), Art. #22, 5 pages. DOI:10.4102/phcfm.v1i1.22

Adekile AD. Haemoglobinopathies. In Azubuike J.C, Nkanginieme KEO, Paediatrics and child health in a tropical region. Owerri. African Educational Services. 1999; 194 - 213

Adeyuyi JO. Knowledge of and attitude to sickle cell disease and sickle carrier screening among new graduates of Nigerian Tertiary Educational Institutions. Postgraduate Medical Journal 2000; 7(3): 120-123.

Adeyemo, Oyenike A.*, Omidiji, Olusesan O. and Shabi, Oluwasola A. African Journal of Biotechnology Vol. 6 (24), pp. 2758-2765, 17 December, 2007.ISSN 1684-5315 © 2007 Academic Journals.

Al-Arrayed S. Campaign to control genetic blood diseases in Bahrain. Community Genet 2005;8:52–55.

Al-Bar MA. Jeddah, KSA: Dar Alnunar; 1991. Malformed fetus, and genetic diseases, etiology, signings, and control.

Alwan A.S, Hamamy H. Riyadh: King Abdulaziz City for Science and Technology; 1993. Hereditary disorders in the Eastern Mediterranean. Arabia. Annals of Saudi Medicine. 1993;13(4):355-9. [PubMed]

Anie K, Smalling B, Fotopoulos C: Groupwork: children and adolescents with sickle cell disease.

Anie KA, Dasgupta T, Ezenduka P, Anarado A, Esuoti I: A cross-cultural study of psychosocial aspects of sickle cell disease in the UK and Nigeria. *Arch Pediatr Adolesc Med* 1994, 148:457-60. PubMed Abstract | Publisher Full Text

Awad H.M. Autosomal recessive osteopetrosis. Annals of Saudi Medicine. 1994;14(2):102–6. [PubMed]

Bazuaye GN and Olayemi EE. Knowledge and attitude of Senior Secondary School Students in Benin City Nigeria to Sickle Cell Disease. World Journal of Medical Sciences 2009; 4(1): 46–49. 7.

Benson RC. Psychologic aspects of gynecologic practice. 5th ed. Beirut, Lebanon: Librairie du liban; 1984. Current obstetric &gynecological

BERNARD & PIERRE, G., Dictionnairemédical pour les régionstropicales, Kangu-Mayumbe BERPS, Congo-Kinshasa1984.

Bhatia M, Walters MC: Hematopoietic cell transplantation for thalassemia and sickle cell disease: past, present and

future.*BMJ* 2008, 337:a1397. PubMed Abstract | Publisher Full Text

Bone Marrow Transplant 2008. 41:109-17. PubMed Abstract | Publisher Full Text

Brown RT, Kaslow NJ, Doepke K, Buchanan I, Eckman J, Baldwin K, Goodman B.Psychosocial and family functioning in children with sickle cell syndrome and their mothers.

Bull World Health Organization 2001, 79(8):704

Cassens B.J. Epidemiology, and prevention of selected acute illnesses, 2nd ed. USA: Williams & Wilkins; 1992. Preventive medicine and public.*Cochrane Database Syst*2002, 3:CD003427. PubMed Abstract | Publisher Full Text

Community Practitioner 2000, 73:556-558.

Deborah Boswell and Rachel Baggaley, VCT and young people, A summary overview, June 2002. DOMART, A. & DOURNEUF, J., Nouveau Larousse médical, Paris, Larousse, 1992.

Diagnosis & treatment.

Erkman J.R. Sickle cell anaemia. Microsoft Student 2007 [DVD]. Redmond, WA: Microsoft Corporation, 2006.

El-Hazmi M. The natural history and the national pre-marital screening program in Saudi Arabia. Saudi Medical Journal 2004;25:1549-54.

Emery A.E.H, Rimoin DL., editors. Principles and practice of medical genetics, vol 1 & 2. Edinburgh: Churchill Livingstone; 1990

Epstein Charles J, Childs B, Clarke FF, McKusick VA, Miller J JR, Motulsky AG, *et al.* Genetic Counselling. *Am J Hum Genet* 1975; 27 : 240-2.

EttieA.Rout, safe marriage: A return to sanity. London, Wood & Sons Ltd, 1922, pp 78, available at http://www.ibiblio.org, accessed on March 26th 2005.

FERRIER, P.-E., Précis de pédiatrie, Paris, Doin, 1984.

Fifty ninth world Health Assembly 2006.

Francis Markus, BBC News, Chinese brides may keep virginity secrets, Many Chinese women are locked in tradition, Wednesday 31 July 2002. www.bbc.co.uk in January 2005.

Gil KM, Wilson J.J, Edens JL: The stability of pain coping strategies in young children adolescents, and adults with sickle cell disease over an 18-month period.

Griffiths, Anthony J. F.; Miller, Jeffrey H.; Suzuki, David T.; Lewontin, Richard C.; Gelbart, eds. (2000). "Genetics and the Organism: Introduction". *An Introduction to Genetic Analysis* (7th ed.). New York: W. H. Freeman. ISBN 0-7167-3520-2.

Gustafson, S. L., Gettig, E. A., Watt-Morse, M., & Krishnamurti, L.(2007). Health Beliefs Among African American Women Regarding Genetic Testing And Counselling For Sickle Cell Disease. Genetics In Medicine, 9, 303-310.

Haggard MP. Hearing screening in children. *Arch Dis Child* 1990;65:1193-5.

Hassan S.P, Hashmi S, Alhassen M, Lawson W, Castro O. Depression in sickle cell disease.

Higgs D. The thalassaemia syndromes. *Q J Med* 1993;86:559-64

Hu Z. Observation on prevention of hepatitis B virus transmission between newly-married couples by HbsAg vaccine. Chung, Hua, Liu, H Hsing

Huisman T. The structure and function of normal and abnormal haemoglobins. In: Higgs D, Weatherall D, editors. The haemoglobinopathies. London: Baillière Tindall, 1993:1-30.

Interdiction mariage pour raison médicale: Indice de fertilité, qualité du mari. available at www.0plus0.com, accessed on February 15th, 2005.

J Am Acad Child Adolesc Psychiatry 1993, 32(3):545-53. PubMed Abstract

J Consult ClinPsychol 1994, 62:856-866. PubMed Abstract | Publisher Full Text

J PediatrPsychol 1995, 20:511-525. PubMed Abstract | Publisher Full Text

J PediatrPsychol 1999, 4:491-8. Publisher Full Text

J.M.Paxman, R.J.Zuckerman, Laws and Policies affecting Adolescent Health, WHO, Geneva. 1987, 300 pages.

Journal of the National Medical Association 2003, 95:533-mnimk

Kabiii I.A 2008. Anthropometric Profiles of Homozygous Sickle Cell Children in North-western Nigeria, *AJOL*, 11, 2.

Kasim A, Adnan A. Risk factors of mental retardation in children attending an educationally subnormal/mental school in Dammam, Saudi

Kliewer W, Lewis H: Family Influences on Coping Processes in Children and Adolescents with Sickle Cell Disease.

Konotey-Ahulu FID: *The sickle cell disease patient.* London: MacMillan; 1991.

Lane PA. Sickle Cell Disease. *Pediatr Clin North Am* 1996;43:639–64.

LANSAC, J., BERGER, MAGNIN, G., Obstétrique pour le praticien, 3ᵉédition, Paris, Masson, 1997.

Laskey, S. L., Williams, J., Pierre-Louis, J., O'Riordan, M., Matthews, A., & Robin, N. H. (2003). Attitudes of African American

Last J.M, Wallace R. Genetic and Public Health. 13th ed. USA: Prentice-Hall International Inc; 1992. Public health & preventive medicine, Mandatory Premarital examinations, available at www.islamtomorrow.com, accessed on November 27th, 2004

London: BailliereTindall 1993;117–50. 20. Thein S. Beta-thalassaemia. In: Higgs D, Weatherall D, editors.

Merger, R., Levy, J., Melchior, J., Précis d'obstétrique, Paris, Masson, 1995: 597 pages

Midence, K., Graham, V., Acheampong, C., & Okuyiga, E. (1994). Increasing awareness for higher quality care. Measuring knowledge of Sickle Cell Disease in Adult Patients. Professional Nurse, 9, 255–258.

Milnnsky A. Baltimore, USA: The John Hopkins University Press; 1992. Hereditary and Your Family's Health.

Moos R.H, Schaefer JA: The crisis of physical illness: An overview and conceptual approach. In *Coping With Physical Illness: New Perspectives. Volume 2.* Edited by Moos RH. New York: Plenum Press; 1984:3-35.

Murphy MF, Wainscoat J, Colvin BJ. Haematological disease. In Kumar P, Clark M, eds. Clinical Medicine. 6th Ed. London. Elsevier Saunders. 2005: 437- 445.

National reproductive health service policy and standards, Ghana National RH Service and Standards, 2nd Edition, Accra, December 2003, 103 pages.

Nigerian Medical Journal 2001, 41(1):21-23.

Nzewi E: Mulevolent oghanje: recurrent reincarnation or sickle cell disease?

Ocheni S. Onah H.E, Ibegulam O.G, Eze M(2007)Pregnancy outcomes in patients with sickle cell disease in enugu, Nigeria. Niger J Med 16:227 – 230.

Ogamdi SO and Onwe F. A pilot study comparing the level of sickle cell disease knowledge in a University in South Eastern Texas and a University in Enugu State, Nigeria. West African Ethn Dis Spring Summer 2000; 10(2): 232-236.

Ohaeri JU, Shokunbi WA, Akindale KS, Dare LO: The psychosocial problems of sickle cell disease sufferers and their methods of coping.

Olsson A. Better prenatal care in Saudi Arabia. Annals of Saudi Medicine. 1985;5(3):169–75.

Olarewaju SO, Enwerem K, Adebimpe WO and Olugbenga-Bello A. Knowledge and attitude of secondary school students in Jos, Nigeria on sickle cell disease. Pan African Medical Journal 2013; 15: 127.

Omonzejele PF: African Concepts of Health, Disease, and Treatment: An Ethical Inquiry.

patients at inner-city neighborhood health centers." *Journal of the National Medical Association*. 98(3): 370-377.

Piel FB, Patil AP, Howes RE, Nyangiri OA, Gething PW, et al. Global distribution of the sickle cell gene and geographical confirmation of the malaria hypothesis. Nat Commun 2010; 1: 104.

Premarital Medical Test May Be Optional, November 2002, available at www.china.org.cn, accessed on 17th January 2005.

Premarital screening, available at www.bnhealthcare.com , accessed on February 10th, 2005

Premarital screening, available at www.bnhealthcare.com, accessed on 16th February 2005.

Psychol Health Med 2007, 12:299-304. PubMed Abstract | Publisher Full Text

PubMed Abstract | Publisher Full Text |PubMed Central Full Text

Rajesh Aggarvall and other specialists medical writers, The Bantam Medical Dictionary,3rdedition,Market House Books Ltd., New York/USA, April 2000:534

Ratip S, Skuse D, Porter J, Wonke B, Yardumian A, Modell B. Psychosocial and clinical burden of thalassaemia intermedia and its implications for prenatal diagnosis. *Arch Dis Child* 1995;72:408–12

Rees DC, Willinms TN, Gladwin MT. Sickle-cell disease. *Lancet* 2010; 376: 2018–31.

Region of World Health Organisation. Proceedings of the symposium on the medical genetics in the setting of middle eastern population

Riddington C, Owusu-Ofori S. Prophylactic antibiotics for preventing pneumococcal infection in children with sickle cell disease.

Ronald, I. Kleinman, M.B., Manuel de planificationfamiliale à l'usage des médecins, London, IPFF. 1981.

Ross PT, Lypson ML, Ursu DC, Everett LA, Rochgues O and Campbell AD. Attitude of Ghanaian women towards genetic testing for sickle cell trait. Inter J Gynae and Obstetrics 2011; 115 (3): 264-268

ROTSART DE HERTAING, I. et J. COURTEJOIE, Maternité et santé, Kinshasa, BERPS, 1988.

Sally Wehmeier, Oxford Advanced Learner's Dictionary of Current English, 6th edition, La Tipographica Varese Italia/Oxford University press, Oxford-England, 2000:1422 p

Serdula M, mark J, Remington P, Ibara C, White M. Premarital rubella screening program: From identification to vaccination of susceptibleSickle cell disorder in the African region. Current situation and the way forward. WHO Regional Committee for Africa AFR/RC56/17-2006.

Soc Sci Med 1995, 40(7):955-60. PubMed Abstract | Publisher Full Text

Soc Sci Med 2001, 52(9): 1403-16. PubMed Abstract | Publisher Full Text

Sylvie L. Cohen & Michele Burger, PARTNERING: A New Approach to Sexual and Reproductive Health, UNPFA, New York, December 2000, 181

Thompson RJ, Armstrong FD, Kronenberger WG, Scott D, McCabe MA, Smith B, Radcliffe J, Colangelo L, Gallagher D, Islam S, Wright E: Family functioning, neurocognitive functioning, and behavior problems in children with sickle cell disease.

Thompson RJ, Gil KM, Keith BR, Gustafson KE, George LK, Kinney TR: Psychological adjustment of children with sickle cell disease: stability and change over a 10-month period.

Uwakwe C.B.U, Kofie SA, Shokumbi WA: Sickle cell disease and the psychological adjustment of sufferers in a Nigerian Teaching Hospital.

Vogel F, Motulsky AG. Human genetics. Berlin: Sprinter Verlag; 1982.

Weatherall DJ, Clegg JB: Inherited haemoglobin disorders: an increasing global health problem.

Weiling, F (1991). "Historical study: Johann Gregor Mendel 1822–1884.". *American journal of medical genetics* 40 (1): 1–25; discussion 26. doi:10.1002/ajmg.1320400103. PMID 1887835.women in the state of Hawaii. Public Health Rep. 1986;101(3):329-33. [PMC free article] [PubMed]

Williams TN, Obaro SK. Sickle cell disease and malaria morbidity: a tale with two tails. *Trends Parasitol* 2011; 27: 315–20.

World Health Organization. 59th World Health assembly.EB117/2006/REC/2. Sickle cell anaemia.[cited 2008 Nov 16].

World Health Organization. Sickle-cell anaemia: Report by the Secretariat. Provisional agenda item 11.4 for the Fifty-Ninth World Health Assembly held on 26th April, 2006. Geneva. WHO. A59/9: 1 - 6.

World Health Organization: Sickle cell Anemia – report by the secretariat. Fifty ninth World Health Assembly 2006.

Yang YM, Cepeda M, Price C, Shah A, Marikad V: Depression in children and adolescents with sickle-cell disease.

Zemel B. S, Kawchack D.A, Ohene-Frempong K, Schall J.I, Stallings VA(2007).Effects of delayed pubertal development,nutritional status and disease severity on longitudinal patterns of growth failure in children with sickle cell disease. Pediatr Res 61:607-613.

Zimmerman, R.K et al. (2006). "Racial differences in beliefs about genetic screening among

APPENDIX

Informed Consent

Dear Respondent,

My name is Adeleke, Adeolu Anthony. I am a Master's degree student of the department of Health Promotion and Education, Faculty of Public Health, College of Medicine, University of Ibadan. I am presently conducting a study titled '*Knowledge, Perceptions and attitude of about to Wed couples towards Premarital Sickle Cell Genetic Testing and Counselling in Ibadan North-East Local Government Registry, Oyo State*'. The purpose of this study is to investigate the level of knowledge, perceptions and attitude towards premarital sickle cell screening.

This questionnaire is designed to gather infotmation concerning knowledge about sickle cell disorder, perceptions, and attitude of about to wed couples towards premarital sickle cell screening in Ibadan North-East Local Government Registry, Oyo state.

The questionnaire was designed based on the ethical principles guiding the use of human participants in research. All information provided shall be kept confidential and in a secured place where no other persons will have access to the information given by the respondents.

Thank you for your cooperation.

QUESTIONNAIRE

Serial number.....

FOR OFFICIAL USE ONLY

Instructions: Please do not write your name.

Please give an appropriate answer by ticking (√) where applicable

I. SOCIO DEMOGRAPHICS:

1. Gender

Male Female

2. Age last birthday less than 20 20 – 25 26– –30 30 and above

3. What is your ethnic origin?

Yoruba Igbo Hausa/Fulani others (specify)

4. What is your current living status?

Alone With spouse/partner with parents with other relatives

With unrelated persons with friends others (specify)

5. Highest level of education Primary Secondary Tertiary

None

6. Religion Christianity Islam Traditional Others (specify)

7. What best describes your current main activity?

In paid employment in self employment Retired Housework Student

job seeking others (please specify).....

II KNOWLEDGE

Please read each question very carefully and then circle one answer that is the best.

Remember, only one answer for each question.

1) Sickle Cell Disease is caused by

a. a virus

b. inherited sickle cell traits from both parents

c. bad blood

d. none of the above

9) Sickle Cell Disease can cause

a. severe debilitating pain

b. strokes

c. infections

d. organ damage

e. all of the above

10) Sickle Cell Disease occurs in

a. boys only

b. girls only

c. white people only

d. black people only

e. all of the above

11) Sickle Cell Pain can feel worse than

a. a broken bone

b. a headache

c. a gunshot wound

d. none of the above

e. all of the above

12) Sickle Cell Disease makes red blood cells

a. round and soft

b. hard and sickle shaped

c. sticky and blue

d. stiff and sickle shaped

e. soft and sickle shaped

13) Sickle Cell Disease is easily managed by

a- antibiotics

b. liver transplant

c. pain relievers

d. blood transfusions

e. none of the above

9) Sickle Cell Disease can cause

a. severe debilitating pain

b. strokes

c. infections

d. organ damage

e. all of the above

10) Sickle Cell Disease occurs in

a. boys only

b. girls only

c. white people only

d. black people only

e. all of the above

11) Sickle Cell Pain can feel worse than

a. a broken bone

b. a headache

c. a gunshot wound

d. none of the above

e. all of the above

12) Sickle Cell Disease makes red blood cells

a. round and soft

b. hard and sickle shaped

c. sticky and blue

d. stiff and sickle shaped

e. soft and sickle shaped

13) Sickle Cell Disease is <u>easily</u> managed by

a. antibiotics

b. liver transplant

c. pain relievers

d. blood transfusions

e. none of the above

14) Sickle Cell Disease could be cured by

a. Bone marrow transplants

b. antibiotics

c. rest

d. none of the above

15) What is the best way of knowing someone carries the gene for sickle cell disease?

a. They look sick

b. They will eventually have Sickle Cell Disease

c. With a simple blood test

d. There is no way of knowing

e. None of the above

16) sickle cell disease can be prevented by

a. Use of condom

b. Premarital genetic testing and counselling

c. Abortion

d. all of the above

e. none of the above

17) What is the best time of testing for sickle cell disease?

a. When feeling sick

b. At any time

c. when one is about to wed

d. all of the above

18) What is the purpose of pre-marital sickle cell testing

a. To know the sickle cell trait status

b. To prevent incidence of sickle cell disorder

c. To make an informed marital choice

d. none of the above

III UTILIZATION OF PREMARITAL SICKLE CELL GENETIC TESTING AND COUNSELLING SERVICES.

19) Do you know that one can check his sickle cell status?

a. Yes b. No

20) Do you know about the availability of sickle cell testing service?

a. Yes b. No

21) Do you know sites where test is done?

a. Family Guidance and Counselling centres	Yes	No
b. Private clinics	Yes	No
c. Government hospitals	Yes	No

d. Others..... (please, specify)

22) The closest distance of the centre to my residence is

Less than 1km 1 – 2km 2 – 3km more than 3km

23). Have you ever done any form of genetic testing and counselling before?

Yes No

24). If yes, was the test free of charge? Yes No

25). If the test was not free, how much did you pay for the test?

less than ₦100 ₦100 – ₦200 ₦200 – ₦300 ₦300 – ₦500 more than ₦500

26). Would you say it is affordable to do the testing? Yes No

27). Was the test time consuming?

too time consuming Time consuming not time consuming

28). Was the test painful?

too painful Painful Not painful

29). What about the providers' attitude?

Friendly harsh not friendly

IV ATTITUDE TOWARDS SICKLE CELL PREMARITAL COUNSELLING AND TESTING

Please tick as appropriate

	Statement	Yes	No	Don't know
30	Do you consider premarital genetic testing important at all cost?			
31	Premarital sickle cell testing and counselling is a waste of time and money.			
32	Premarital sickle cell testing and counselling should be made mandatory.			
33	Marriage institutions should encourage their members to go for pre marital sickle cell testing and counselling.			
34	I would encourage my partner to be tested for the sickle cell trait if I was found to be a trait carrier.			

V QUESTION ON PERCEPTION OF SICKLE CELL DISORDER AND HEALTH BELIEFS

Please rate your level of agreement

Perception	Strongly agree	undecided	Strongly disagree
Severity			
35) Sickle Cell Disease is a serious disease			
36) Having a Child with SCD would be very scary			
37) My life would change if my child had SCD			
38)Children with sickle cell disease are at risk for infections, Pain, pneumonia and stroke			
Susceptibility			
39) If both partners are carriers of sickle cell trait, children could be at risk for Sickle Cell Disorder.			

40)Sickle Cell Trait is transmittable to offspring			
Sickle cell disorder is only preventable if only one of the partners has sickle cell trait.			
41)Anyone may be a carrier of sickle cell trait			
Benefits			
42) Sickle Cell Disorder can be prevented in offspring if both partners know their sickle cell trait status.			
43) Knowing my sickle cell trait status will make me confident in forming relationships.			
44) Knowing the burden of having a child with SCD would make me engage in pre-marital sickle cell testing and counselling service.			
Barriers			
45) Testing for sickle cell trait is painful and difficult			
46) My partner would be hard to convince to have testing			
47) I would not want to pay for sickle cell trait testing because it is expensive.			
48) I am worried that the results could get into outsider's hands.			
49) Genetic testing is not acceptable because the results may lead to discrimination against sickle cell trait carriers.			
50) Service centres are distant to my residence.			
51) My culture does not support premarital sickle cell testing and counselling.			
52) Genetic testing is not acceptable by my faith			

DRAFT IN-DEPTH INTERVIEW GUIDE ON KNOWLEDGE, PERCEPTIONS AND ATTITUDE OF ABOUT TO WED COUPLES TOWARDS PREMARITAL SICKLE CELL GENETIC TESTING AND COUNSELLING IN IBADAN NORTH-EAST LOCAL GOVERNMENT REGISTRY IN IBADAN NORTH- EAST LOCAL GOVERNMENT REGISTRY, OYO STATE

Good day Sir, My name is.....,this study is being conducted as part of requirements for the award of (MPH) Degree of Department of Health Promotion and Education in the Faculty of Public Health, University of Ibadan.

The study is designed to investigate *the Knowledge, Perceptions and attitude of about to Wed couples towards Premarital Sickle Cell Genetic Testing and Counselling in Ibadan North-East Local Government Registry Local Government Registry, Oyo State.* Your participation in this interview is to enable me understand your knowledge, perceptions and attitude towards topic.

Please note that your patticipation in this interview is voluntary, hence you may agree or decline to participate. There are no wrong or right answers, all responses are very important to this study. I also seek your permission to capture this interview on a recorder, to ensure no part of this interview is lost during analysis

Thank you.

Would you like to participate? Yes No

ADELEKE ADEOLU
08036169412

S/N	QUESTIONS	PROBE
1	What is your view about sickle cell disorder?	• Can you really describe it?
2	What do you think can cause sickle cell disorder?	• Does it affect only the child or the entire family? • Can it really be prevented? • What effort should be put in place to prevent it?
3	Management options?	• Do you know of management options for SCD? • Is it cheap or expensive?

		• Does it have a cure? • Is it really easy to get the cure? • Are you aware of a successful cure in Nigeria?
4	Should Premarital Sickle Cell Counselling and Testing (PSCCT) be made mandatory?	• What is your take about PSCCT? • Should every marriage institution enforce or encourage it? • Do you know of facilities where one can do the testing? • Do you think service centres will be accessible by people? • Do you think it is expensive to do PSCCT?
5	Do you think PSCCT is beneficial?	• How enthusiastic do you think people are about PSCCT? • DO you think a sickle cell carrier should continue a relationship with another carrier even if discovered?

MINISTRY OF HEALTH

DEPARTMENT OF PLANNING, RESEARCH & STATISTICS DIVISION

PRIVATE MAIL BAG NO. 5027, OYO STATE OF NIGERIA

Your Ref. No. .....

All communications should be addressed to the Honorable Commissioner quoting

Our Ref. No. AD 13/ 479/___

April, 2015

The Principal Investigator,
Department of Health Promotion and Education,
Faculty of Health,
College of Medicine,
University of Ibadan,
Ibadan.

Attention: Adeleke Adeolu

Ethical Approval for the Implementation of your Research Proposal in Oyo State

This acknowledges the receipt of the corrected version of your Research Proposal titled: "Knowledge, Perceptions and Attitude Sickle cell Genetic Testing and Counselling in Ibadan North-East Local Government Registry, Oyo State."

2. The committee has noted your compliance with all the ethical concerns raised in the initial review of the proposal. In the light of this, I am pleased to convey to you the approval of committee for the implementation of the Research Proposal in Oyo State, Nigeria.

3. Please note that the committee will monitor closely and follow up the implementation of the research study. However, the Ministry of Health would like to have a copy of the results and conclusions of the findings as this will help in policy making in the health sector.

4. Wishing you all the best.

OYO STATE RESEARCH ETHICAL COMMITTEE

Signature & Date

Solagbade (Dr)
Director, Planning, Research & Statistics
Secretary, Oyo State, Research Ethical Review Committee

MINISTRY OF HEALTH

DEPARTMENT OF PLANNING, RESEARCH & STATISTICS DIVISION

PRIVATE MAIL BAG NO. 5027, OYO STATE OF NIGERIA

Your Ref. No. .....

All communications should be addressed to the Honorable Commissioner quoting

Our Ref. No. AD 13/ 479/___

April, 2015

The Principal Investigator,
Department of Health Promotion and Education,
Faculty of Health,
College of Medicine,
University of Ibadan,
Ibadan.

Attention: Adeleke Adeolu

Ethical Approval for the Implementation of your Research Proposal in Oyo State

This acknowledges the receipt of the corrected version of your Research Proposal titled: "Knowledge, Perceptions and Attitude Sickle cell Genetic Testing and Counselling in Ibadan North-East Local Government Registry, Oyo State."

2. The committee has noted your compliance with all the ethical concerns raised in the initial review of the proposal. In the light of this, I am pleased to convey to you the approval of committee for the implementation of the Research Proposal in Oyo State, Nigeria.

3. Please note that the committee will monitor closely and follow up the implementation of the research study. However, the Ministry of Health would like to have a copy of the results and conclusions of the findings as this will help in policy making in the health sector.

4. Wishing you all the best.

OYO STATE RESEARCH ETHICAL REVIEW COMMITTEE

Signature & Date

Solagbade (Dr)
Director, Planning, Research & Statistics
Secretary, Oyo State, Research Ethical Review Committee