Health Information Management (HIM) System as a Positive Protagonist for Genetic Diseases in Nigeria (A case study of Sickle Cell Disease (SCD) Patients)

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ABSTRACT: *Sickle Cell Disease (SCD) which comes in diverse ranges of forms of Hemoglobin disorders (such as sickle cell hemoglobin S-HbSS and hemoglobin C-HbSC, sickle cell anemia (SCA), beta and alpha thalassemias, SD, and SE, etc.). SCD is a common genetic disorder in most sub-Saharan African countries with affecting rate of up to three percent of parturitions (births) as witness in various parts of the continents and especially in Nigeria. It is one of the most uncared non-communicable genetic disorder diseases that are characterized by series of anemia and sickling of red blood cells (RBCs) among Nigerians in recent time. The most common form of SC disorder is produced from the homozygosis of beta-globin S gene mutation (HbSS disease). These red blood cells disorders are portrayed with series of chronic, hemolytic anemia, various injury, vaso-occlusive crisis, stroke, and organ dysfunction in some cases. Patients (which includes children and adults) with SCD are familiar with relate traits and indications, problems and complications of the disease which increases with age. There have been reports of patients' addiction to different types of prophylaxis drugs of SCD such as Tramadol, Pentazocine (Fortwin), Diclofenac injections, etc.; which are prevalent in tertiary institutions. The adequacy of proper data and information management support for managing SCD can reduce the rate of high early-mortality which has been on the rise in children, young adults and adults in Nigeria. The addition of health information management (HIM) programs for the SCD among these pairs will create an affordable method to perform an evaluation of SCD mortality among Nigerians with sufficient health information management.*

Key Words: *Sickle Cell Disease (SCD), Health Information Management, Hemoglobin, Thalassemia*

I.INTRODUCTION

Sickle Cell Disease (SCD) disorder is one of the global health challenges with series of implications which ranges from little to psycho-social levels. In Nigeria today, SCD disorder is one of the most prevalent genetic red blood cells (RBC) disorder affecting citizens of the country of diverse age groups resulting to hemolysis and vaso-occlusive crisis. It was reported that Nigeria has the largest population of people living with SCD disorder with about 150,000 parturitions (births) annually. This survey has also shown that people living with SCD have abnormality in their RBC with their hemoglobin referred to as *hemoglobin S* or *sickle hemoglobin* as featured in their RBC. Human RBC is characterized with this hemoglobin which is the protein that carries oxygen throughout the body system. SCD as a genetic RBC disorder is a noncommunicable disease as well as a disease that is transferred through gene from parents to their offspring. It is not communicable like other transmittable diseases like Ebola, Zika, Tuberculosis or common ailments like cold or flu. Patients living with SCD undergo various variability of the disease with different signs and impediments (complications) such as several years of acute pain from birth (i.e. vasoocclusive episodes (VOEs)), long-lasting pain, several multiple organ injury (e.g. kidney failure, heart failure, etc.), short lifespan, bone infections, stroke among others [1]–[4]. The high frequency of sickle cell anemia disease in Nigeria is one of the highest in the world with an approximation of over 100,000 Nigerian children being given birth to each year with the SC disorder [5]. Patients with this disorder writhe more than average with high frequency of illness and premature deaths especially in children and this can be aggravated to severe attacks such as heart attacks, kidney failure severe bone infections and sudden mortalities in patients that survive the infancy stage of attacks [5].

In a country of a population estimated to be over 160 million people, it has been statistically shown that over 41 million citizens are carriers of the *'S'* gene of which this approximations total is far above the total number of victims of this carrier in other countries altogether in Africa. In spite of this large statistics of citizens suffering from the SCD disorder or carriers of it, the Nigerian populace continue to demonstrate an uncared attitude to SCD patients and not only this, the general public

has a negative perceptions and wrong health information management defiance towards it [5].

National Institute of Health (NIH) in [6] on the SCD disorder demonstrated that people living with SCD disorder genetically inherits two abnormal hemoglobin genes (one from each parent). Generally from research, it has been researched that for all traits of SCD, no less than one of the two abnormal genes causes a person's blood to produced hemoglobin S. In cases where a person has two hemoglobin S genes resulting to the *Hemoglobin SS*, the resulting disease is referred to as *Sickle Cell Anemia (SCA)*. This is the greatest collective and frequently severe type of SCD disorder prevalent in Nigeria. Other forms of SCD observed in Nigeria of recent are Hemoglobin SC disease and hemoglobin S thalassemias (hemoglobin Sα thalassemia, hemoglobin $S\beta^0$ thalassemia, $S\beta^+$ thalassemia, hemoglobin SD, and hemoglobin SE.

In advanced and developed countries like US, UK, Canada and European countries where the SCD disorder is not rampart, the mean life expectancy of individuals living with the homozygous sickle hemoglobin mutation (HbSS) is estimated at 40-60 years and which has improved over the last few decades due to proper health information management [1]. This success development witness an overall increase in Paediatric survival of patients with SCD with the introduction of Prophylactic Penicillin Study carried out in 1986. The discovery demonstrated that the prescription and usage of prophylactic penicillin could alleviate life threatening infections in affected children. Hence, from the study, it was also discovered that collective newborn babies screening became a standard practice especially in US in late '90s and in UK in early 2000 which enabled early diagnosis and patient management [1]. In another development to support proper health information management, introduction of pneumococcal conjugate vaccine which significantly contribute to the decrease in SCD mortality in children younger than 10 years of age were also observed as recorded in [1]. Unfortunately, in less and under developed countries like Nigeria, averafely 50% of children younger than 5 years of age die due to misplaced health diagnosis, complication and non-proper health information management of SCD [1], [2], [7].

Subsequently, more than 98% of children with insignificant or trivial traits of SCD in developed countries are growing and living into adulthood, SCD has become a lingering disorder requiring proper comprehensive life-long management especially in Nigeria. The major challenges of this life threatening disorder facing the adult population in the country include struggles managing the transition from paediatric care into adult care due to lack of available healthcare information management and proper healthcare provision for young adults and adults with SCD. Hence, youths and adults living with SCD disorder depends solely on Emergency Departments (EDs), medical doctors (hematologist), inpatients and outpatients treatment sessions for their care and in some cases these are not available. Improvements have been witness of late in some of the country's University Teaching Hospitals with the emergence of Hematology Departments (HD) to oversee the prevalent scourge of SCD disorder. Therefore the purpose of this paper is to awaken the primary health care medical doctors, hematologists, inpatients and outpatients' doctors, home cares, ED medical doctors and the government with the up-to-date understanding and proper health information management of the SCD through Information and Communication Technology (ICT).

II.SYNOPSIS OF SICKLE CELL DISEASE

From medical biology point of view, a *cell* often called the *'building block of life'* is the smallest unit of life that can replicate independently. It consists of cytoplasm enclosed within a membrane containing different biomolecules such as proteins and nucleic acids. Various organisms can be classified either as a *unicellular* (single cell) or *multicellular* organisms (including plants, animals and mammals). While the number of cells in plants and animals varies from species to species, human body contains more than 10 trillion different cells. These cells worked together to sustain life with the support of other non-cellular components of the body such as water, macronutrients (e.g. carbohydrates, proteins, lipids), micronutrients (e.g. vitamins, minerals) and electrolytes. Tissues, on the other hand, are collections of human cells that functioned together in order to perform one single task. Multitudes of tissues function cooperatively to produce an organ that completes specific functions in human body. Despite this structural organization, the entire activities within human body continuously depend solely on the cell. In another perspective, it is a complex unit that makes life possible [1], [6].

Organelles, an important human body component, are the structural component that permits human body to maintain life. They are suspended within a gelatinous matrix called the *cytoplasm* which is controlled within the cell membrane of human body. One of the few cells in the human body that

lacks almost all organelles is the Red Blood Cells (RBC). The functions of different cells within human body vary basically on either the type or location of the cells in the human body. The organelles function together to preserve the cell alive and allow it to carry out its specific purpose. Sometimes these organelles are highly dedicated and can vary in size, shape and number. Nevertheless, even though these organelles are utmost functional units, they can neither single handedly exist nor operate alone without the presence of the cell as a whole.

Therefore, in human body, tissue cells require constant supply of oxygen to perform very well. As such, the hemoglobin RBC normally takes up oxygen from the lungs and carries it to the tissues within the body. RBCs containing normal hemoglobin are disc-shaped in nature (doughnut-like shape without a hole), which allows the cells to be flexible for easy movement through large and small blood vessels for the purpose of delivering oxygen. On the other hand, the sickled hemoglobin is not like the normal hemoglobin as it develops stiff rods within the RBCs, changing it into a semicircular or sickle shape. The sickle shaped cells developed are not flexible and as such they stick to the vessel walls of the human cell, causing a blockage that slows or stops flow of blood thereby initiating shortage or total obstruction of oxygen to reach nearby tissues [6] as depicted in Figure 1 (*a*) and (*b*).

The absence of adequate oxygen in tissue cells can initiate series of attacks from mild to unexpected severe pain known as *sickle cell crisis* or *pain crisis*. The crisis attacks can transpire unnoticed and unannounced to the patients and this prompts patients with SCD to visit clinics for effective checkups and treatments. Records have also shown it that most children genetically born with SCD disorder observed pain free occurrences amid painful crisis but this is predominant in young adults and adults with long-lasting enduring discomfort. The presence of sickling RBCs and the absence of adequate oxygen delivery into the tissue cells can results into human body organ damage. Complications such as injury to patient's heart, brain, spleen, eyes, liver, joints, bones, marrow, kidneys, lungs, penis, and skin appear in SCD patients over a lifetime [5], [6], [8]

The resulting inflexible sickle shaped cells form due to lack of oxygen to the tissues can't reshaped easily to the normal flexible type; they tend to burst apart (hemolysis). Normal RBCs have a 90-120 days life span while the sickle cells usually live not more than 20 days (precisely between 10-20 days). Human body always make new RBCs in order to replace damaged old cells due to illness or diseases, as such in SCD patients, the body finds it difficult in renewing these damaged cells, in this case, the number of RBCs is frequently lower than normal resulting into what is known as *anemia* (having less energy) [1], [5]–[7], [9], [10]

Records has shown it that SCD is a life-long disease and its severity varies widely from patients to patients and as such having the right health information and managing the disease well will reduce the high rate of mortality in under-developed countries. Rapid improvements has been witness in developed countries such as US, UK, Canada, etc., witnessing a high life expectancy from 14 years to a range of 40-85 years for SCD patients. Presently, one of the appropriate discovered therapy for SCD patients is the Hematopoietic Stem Cell

Transplantation (HSCT), but it is likely unfortunate that most of the patients that can benefit from these cure are either too old for the therapy or could not have a willing and good genetic matched donor as a relative to stand in as a donor for the transplant. For a successful HSCT process, a well matched donor is needed and expected to proffer the best chances for a successful HSCT process. For Nigeria, where the rate of donors are very low or even absent, effective management and treatments can help in reducing the symptoms or crisis and prolong the life of the patients. In another perspective, early diagnosis through proper health information and constant medical care can avert or reduce complications thereby contributing an enhanced healthcare and comfort of the SCD patients [1]–[7], [9], [10]

SCD disorder comes from the output of a single-point transformation (mutation – the replacement of glutamic acid with valine in position 6 on the beta globin (β-globin) subunit of hemoglobin) [1], thereby causing deformation or transmutation of hemoglobin known as *sickle hemoglobin (HbS)*. Patients that genetically inherit two copies of these HbS transmuted or mutations are known as *homozygous (HbSS)* possessing the phenotype form of the disease while those who inherit just one copy of the HbS mutation are called *heterozygous carriers (HbAS)* as they do not exhibit the clinical diagnose disease traits called the *sickle cell trait*. In other words, other types of SCD disorder occurs when mutations responsible for anomalous types of hemoglobin either "*C*" or "*E*" or β-thalassemia combines with HbS as a compound heterozygous mutations producing hemoglobin genotypes such as SC, SE, $S\beta^0$, or $S\beta^+$. It has been noted and reported that people living with HbSS and HbSC are common in Nigeria and

also have the greatest unbearable forms of SCD disorder [1], [2], [6], [7], [11].

Abnormal hemoglobin known as *hemoglobin S* results into all the complications experience by a SCD patient. The problem with hemoglobin S is caused by minor deformation in the gene which produces the beta globin (Sβ globin) which changes the way the hemoglobin works [7]. Figure 1 shows a

pictorial image of both the normal and abnormal RBCs hemoglobin. The figure also depicts the shapes of the normal and the sickled RBCs hemoglobin [6]

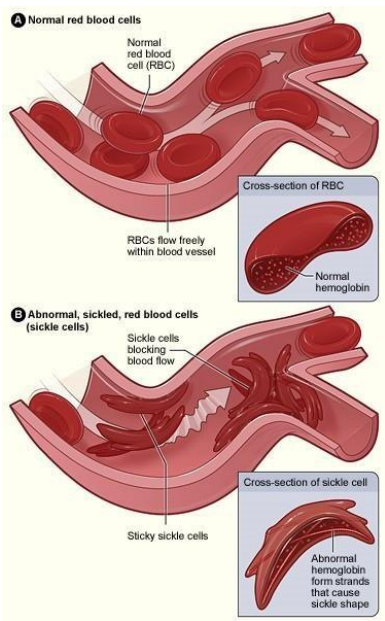

Fig. 1.(a.) Normal RBCs flowing freely showing normal hemoglobin (b.) Abnormal sickled RBCs blocking blood flow in blood vessels showing crosssection of sickled cell with abnormality forming irregular stiff rods [6]

III.GENETICS PATTERN OF SCD

Generally, from medical biology point of view, normal human beings have the following hemoglobin in their bodies. They are:

- hemoglobin *A*: consists of two alpha (α) and two beta (β) chains,
- hemoglobin *A2*: consist of two alpha (α) and two delta (δ) chains and
- hemoglobin*F*: consist of two alpha and two gamma (γ) chains.

From these points, hemoglobin *F* controls the early years of childhood until 6 weeks of age while hemoglobin *A* governs human beings throughout their life time [5], [6].

With this analogy, their sickle-cell conditions in the case of SCD patients possess an autosomal recessive pattern of inheritance from their parents. In this vein, the hemoglobin type a patient inherits from the RBCs depends solely on what type of hemoglobin gene are genetically inherited from their parents. Where one of the parent exhibits sickle cell anemia traits and his or her spouse also exhibits the same sickle cell traits, there is a 50% probability of the person giving birth to a child having a SCD disorder and a 50% chance of having a symptoms or exhibit the sickle cell trait as a carrier. On the other hand, when both parents have a SC trait, the dependents have a 25% chance of SCD, 25% of not carrying the SC alleles but a 50% chance of exhibiting the heterozygous disorder as shown in Figure 2 [1][5][6].

Furthermore, in the case where hemoglobin S gene is inherited from only one parent and a normal hemoglobin gene is inherited from the other parent, there is a tendency that the offspring will have the sickle cell trait as a carrier of the sickle hemoglobin. Children born out of this combination are always healthy; only rarely do people with SC trait have complications similar to those seen in patients with SCD disorder. Nevertheless, people with SC trait are carriers of a defective hemoglobin S gene; hence, there is a probability chance that they can pass it on to their children when they gave birth. On the other hand, in the event that the child's other parent also has sickle cell trait or another abnormal hemoglobin gene like thalassemia, hemoglobin C, hemoglobin D, hemoglobin E, then that child has a chance of having a sickle cell disease disorder.

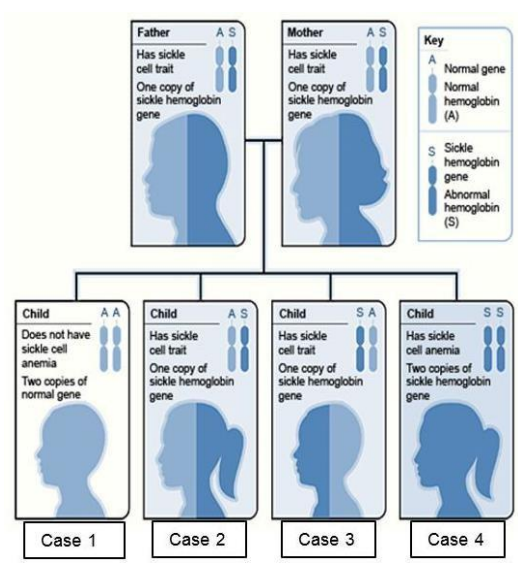

Fig. 2.SCD Inheritance Pattern [6]

SC genes mutation possibly arise naturally in different geographical zones as recorded and proposed by restriction of endonucleic analysis as

reported by [12] and [13]. These alternatives country areas such as Benin, Cameroun, Bantu, Saudi Arabia and Senegal has recorded clinical importance because majority of them are connected with higher HbF levels especially in Senegal and Saudi-Asian and they also have a tendency to suffer milder complication of the SCD disorder [12]. However, patients that are heterozygous in nature for HbAS (carriers of sickling hemoglobin), the bonding problems are minor because the normal allele is able to produce over 50% of the hemoglobin. On the contrary, patients exhibiting the homozygous HbSS, the presence of long chain of polymers of HbS distort the shape of the RBC from a smooth doughnut-like shape of the normal RBC to shabby and full points sickled abnormal shapes (as shown in Figure 1(b), rendering them brittle and vulnerable to breaking within the capillaries [5]. SC trait carriers exhibit symptoms of sickling patients only in the event they are stressed up, or their tissue cells are deprived of oxygen (hypoxia), acidosis (increase in acid content in the blood) or severely dehydrated [1][5]. The sickle cell disease crisis follows when the sixth amino acid, called the glutamic acid, is replaced by the valine by changing its structure and function. In another perspective, another name given to sickle cell anemia is E6V. Valine, which is hydrophobic in nature, initiates the hemoglobin to collapse on itself frequently but the arrangement remains the same. Therefore, the numerable amount of collapsing of the hemoglobin on itself causes the RBCs to become sickle in shape [1], [6]. The mutation of single nucleotide of β-globin gene is the gene defects which causes the replacement of the glutamic acid by the valine at position 6 of the hemoglobin chart [1], [3].

The alteration (mutation) caused by the hemoglobin S is known as *HbS* compared to the normal RBCs hemoglobin called the *HbA*. The hydrophobic side chain of the valine filtrate at position 6 of beta chain is associated with the hydrophobic patch resulting in hemoglobin S molecules to aggregate and form fibroid precipitation in some humans. The allele responsible for sickle cell anemia can be found on the short arm of chromosomes 11 (i.e. 11p15.5) as recorded by [1] and [6]. SCD patients obtains the defective gene from both parents that produces a copy of sickle hemoglobin gene as shown in Figure 2, while patients who receives one defective and one healthy allele remains healthy, but ironically they can pass the disease onto their descendants, who are referred to sickle cell disease (SCD) carriers or heterozygote. However, heterozygotes also are very vulnerable to malaria disease but they experience less severe symptoms of SCD disorder traits

[5], [6].

Owing to a very good adaptation benefit of the heterozygote, the SCD disorder is predominant common especially among citizens of the descendants of the lineage of prominent African countries in malaria-troubled areas. Malaria parasites in Nigeria have a complex lifecycle and spend part of its cycle in RBCs. For sickle cell carriers (i.e. *HbAS*), the presence of the malaria parasite in the RBCs of the sickle cell patients causes the defective RBCs hemoglobin to rupture precipitately thereby causing the malaria *Plasmodium parasite* unable to reproduce. Moreover, the process of bonding hemoglobin affects the ability of the malaria parasite to digest the hemoglobin in the first place. Therefore, in areas where malaria is predominantly a common and rampart disease, patients with SCD disorder has a slim chance of survival if they are infected with the SC trait and at the same time attacked with malaria parasite especially the *P. falciparum*. As such, it is important in malaria endemic countries like Nigeria, patients with sickle cell anemia traits (SCA) and particularly children must be protected from malaria by taking necessary and suitable prophylaxis. In less endemic malaria areas like the US, UK, the occurrence of SC anemia among citizens especially African-Americans is lesser (approximately 0.25%) than in Nigeria which is about 4.0% and still declining. With the absence of endemic malaria in high profile zones, sickle cell mutation is honourably not celebrated and tends to reduce in the affected zones like Nigeria by process of natural selection and presently through artificial prenatal genetic screening as observed in US and other developed countries

[1], [2], [5], [6], [11]

IV.PHYSIOPATHOLOGY AND MEDICAL INDICATORS OF SCD

RBC elasticity damage is paramount to physiopathology of sickle cell disease (SCD) disorder. Regular or ordinary RBCs are relatively elastic in nature and this gives the tissue cells to buckle or bend in order to penetrate through the human body capillaries as shown in Figure 1. In sickle cell disease patient, inadequate oxygen tension increases the tendency of the RBC sickling thereby the repeated sickling occurrence damage the membrane thereby causing a decrease in tissue cells elasticity. Because of these, the cells find it difficult to reshape to the original or natural shape when the tension in the tissues is finally restored through introduction of oxygen. Therefore, these rigid RBCs finds it difficult to

buckle as they penetrate through the narrow passage of the capillaries leading to vessel occlusion (vaso-occlusion in SCD patient) and isochaemia as depicted in Figure 3 [1][6]. The actual SC anemia morbidity is caused by hemolysis (annihilation of RBCs) due to their sickled shape. Even though bone marrows endeavours to compensate by recreating new RBCs, this cannot be compared to the high level of damage that has already been done [6].

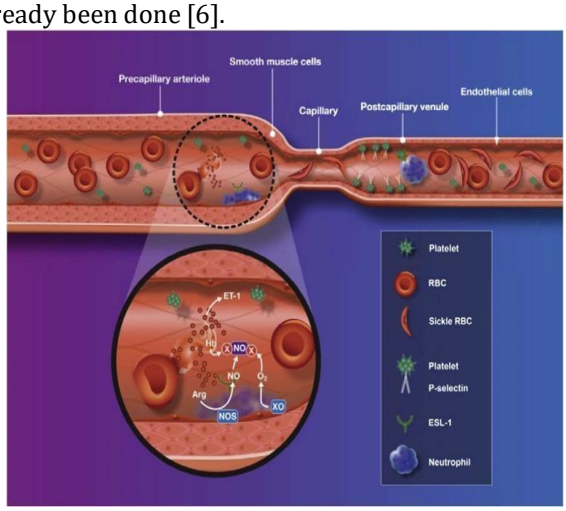

Fig. 3.Physiopathology of SCD vaso-occlusion (Keys: Arg (arginine); ESL1 (E-selection ligand-1); ET-1 (endothelin-1); Hb (hemoglobin); NO (nitric oxide); NOS (nitric oxide synthase); O2– (superoxide); RBC (red blood cell); XO (xanthine oxidase)) [1], [5]

Medical indicators are very important in SCD management where the sickle cell disorder shows phenotype heterogeneity which results from both genetic and environmental influences. For example, in HbSS, the complete blood counts (CBC) and packed cell volume (PCV) are very vital. Research has shown that CBC reveals hemoglobin levels in the range of 6-8g/dl with high reticulocyte count when the bone marrow tends to recompense for sickled cells destruction by producing more RBCs while PCV on the other hand has its range for male above 18 years of age from 40-52% and female 35-47% in normal hemoglobin. The ranges for the PCV for SCD patients might change alternatively in a decline value with respect to both male and female. However, in other forms of SCD, Hb level are likely to be higher [1], [14]. The abnormality of the hemoglobin can be detected through hemoglobin electrophoresis test (blood test perform to check the different types of hemoglobin present in the blood). Its results are usually ready within several days where various types of hemoglobin move at varying speeds. The sickle cell hemoglobin (HbSS) and hemoglobin C with sickling (HbSC) are major common disorder forms in Nigeria and are detected easily through this test.

One of the medical indicators common to SCD patients in Nigeria is acute or chronic pains which the trademark of SCD disorder. It can arise out of small vessels blockage or constriction and subsequent tissue infarction (death of tissues due to absence of oxygen), organ damages, or be idiopathic (occurring without known cause). Vaso-occlusive episodes (VOEs) are painful occurrences that occur from vasoocclusive crisis with inflammatory and ischemia (restriction of blood supply to tissues) consequences. These VOEs indicators can arise throughout the body of the patient from bones to muscles, mesentery and other organs [1], [14]. Acute sickle cell crisis often precipitate by infections where medical test such as urinalysis (detecting occult urinary tract infection), chest X-ray (occult pneumonia) should be regularly conducted in order to guide against it. In SCD crisis, chronic pain which is poorly understood and managed in Nigeria can be devastating and often times resulted into terrible complications such as leg ulcers, avascular necrosis (death cells due to enzyme degradation), and or neuropathic complications [1].

Acute or chronic pains are not the only medical indicators for SCD disorder. Other defined SCD crisis include aplastic (reduced function of bone marrow), hemolytic crisis (rapid increase in blood cells failure causing PCV level to drop), sequestration (blood pooling in organs), acute chest syndrome (ACS) and stroke (a common recent phenomenon in some patients in Nigeria especially young adults and children) [1], [3], [4], [6], [10], [11], [15][16]. Table 1gives an illustration of major medical indicators of children and adults with SCD and their various expected symptoms.

Genetic counseling is usually rendered to couples planning to get married and have healthy children especially to those that are carriers of the sickle cell disease disorder. For proper, positive and effective counseling, a test is conducted to find out if an unborn child has the disease in the case of pregnant women that are carriers. This is done by either taking blood samples from the fetus or samples of amniotic fluid (pregnant woman's water). However, it has been reported that taking blood samples from the fetus has greater risks than from the amniotic fluid; hence, the latter test is usually used. In addition

Sickle cell disease disorder patients have an increase tendency of being infected and affected by diseases and also they are susceptible to high risk of various life threatening complications such as acute chest pain syndrome, stroke, multi-organ

wounds and subsequent end-organ damage, sepsis, pulmonary hypertension, hepatic disease, pulmonary embolism (blockage of an artery in lungs) and cardiomyopathy, etc. The greatest threats of patients living with sickle cell disease disorders in Nigeria are reduced lifespan, inadequate quality of life due to inadequate health management, high rate of anxiety, poverty, lack of health facilities in rural areas, additions to various prophylaxis, and finally high rate of mortality of patients due to lack of proper healthcare [1], [5]–[7], [10], [14]

TABLE I. MEDICAL INDICATORS OF CHILDREN AND ADULT SCD(ADAPTED FROM [1])

Children with SCD			Adult with SCD showing additional indications and problems
Indications/Symptoms			
	Infants	Children	
	Pain in chest, abdomen, and limbs/joints	Pain (acute or chronic)	Severe joint pain
	Dactylitis	Acute anemia	Chronic leg ulcers
	Anemia	Infections	Retinopathy
	Mild jaundice	Jaundice	Thromboembolic complications
	Enlarged spleen	Poor nutritional status and growth	Neurocognitive impairments
	Fever	Academic failure	Narcotic dependence/tolerance
	Frequent upper respiratory infections	Delayed puberty	
Complications			
CNS	Stroke		Recurrent ischemic stroke, hemorrhagic stroke
Eye	Retinal artery occlusion/retinopathy		Progressive retinopathy
Lung	ACS		Recurrent ACS
	Asthma		Pulmonary hypertension
			Chronic lung disease
			Premature coronary artery disease
Heart	Left ventricular hypertrophy		Heart failure
	Cardiomyopathy		Auto-infarction
Spleen	Acute splenic sequestration		Functional asplaenia
	Impaired immunity (e.g. bacterial infection, sepsis)		
Liver			Hepatic sequestration
			Liver failure secondary to transfusional iron overload
Kidney			Nephropathy
	Hyposthenia		Frequent urinary tract infections
	Proteinuria		
	Renal impairment/failure		
Gall bladder	Cholelithiasis		Cholelithiasis
Genitals	Priapism		Priapism
Bones/joints	Avascular necrosis		Avascular necrosis
	Aplastic crisis		Early loss of bone density Chronic leg ulcers
Skin	Chronic ulcers, typically on the ankles		

Keys: CNS (Central Nervous System), ACS (Acute Chest Syndrome).

V.SIGNS,SYMPTOMS AND COMPLICATIONS OF SCD

SCD patients show early signs of the disorder from infants. Every sickle cell disease disorder patients has the disease right from childhood but most children do not have any problem related to the disease until about 5-6 months old while some 8 months. As such, developed countries like US have mandated that all newborn babies receive screening for SCD disorder and parents of SCD patients are notified on time before the child has the symptoms. Some patients show their own sign later in life. In Nigeria, in the past, there has been no stable and adequate programs like the developed countries on managing the SCD disorder early enough especially in the rural areas of the countries but of recent awareness and sensitization has been going on with the advent of hematology departments in various clinics and teaching hospitals in the country. Various foundations and small clubs in the urban areas are trying their best to make sure these process of health information and management is done early enough but the process is not enough or very slow as it pose high risk to the patients and as such increase the rate of mortality [1], [3], [5].

Children or infants exhibiting these sickle cell disease disorder will begin to experience traits of sickle cell problems in their early age days on and symptoms of SCD may include the following among others dactylitis (painful swelling of the hands and legs), anemia fatigue or fussiness, mild jaundice (yellowish color of the skin, nails or eyes), icteris (eyes whitening – occurs when a large number of RBCs hemolyze), enlarged spleen, fever, frequent upper respiratory infection, and so on. These sickle cell disease disorder signs and symptoms varies from one individual to another and can change over time, nevertheless most of them are related to complications of the SCD disorder [5], [6].

In another way, SCD disorder may lead to various acute long-lasting complications and have high rate of mortality [5]. The major complications

predominant in Nigeria of recent and most reported ones are as follows which therefore falls in the purpose of this paper which is to awaken the primary health care physicians, hematologist, inpatients and outpatients doctors, home cares guardians to oversee the prevalent scourge of SCD disorder in the country.

A.Vaso-Occlusive Episodes (VOEs)

VOEs crisis is one of the various complications of sickle cell disease disorder caused by sickle-shaped RBCs that hinders the capillaries and restrict body organs blood flow resulting to ischemia, acute pains, necrosis, and sometimes damage to the organs. The incidence, severity and duration of these crises vary significantly from one individual to another. Painful crisis are treated or managed with proper dehydration, correct prescribed analgesics, and in some cases blood transfusions. Appropriate opioid supervision being monitored at regular interval during the crisis until it subsided should be given all through the pain management period. A subgroup of patients' management routine for Non-Steroidal AntiInflammatory Drugs (NSAIDS) such as diclofenac, naproxen, Pentazocine (fortwin) can be administered during minor crisis. The later of the NSAIDS (Pentazocine) has been reported of recent to be adversely abused by young adults as it has the same symptoms and components as other narcotic drugs especially for relief in severe pains [5], [6]. VOEs crisis can strike almost anywhere in the body and in more than one point or place of the human body at a time. But often various places in human body that has been recorded where the pain occurs are the lower back, legs, arms, abdomen, chest, and of recent in the brain. These crises can be initiated by illness e.g. fever (prominent), weather/temperature changes, stress, dehydration (not drinking enough water), or at high altitudes. However, in most cases, the patients does not know what triggers or causes the crisis [5], [6], [11], [15], [17]. Young adults and adults living with SCD disorder suffer various complications from chronic pain and it has been shown to be a tough time for individuals to describe these experiences but it is usually different from ordinary pain that occurs from other damaged organ. Some of the VOEs and long-lasting pain if not well managed can lead to high mortality.

B.Severe Anemia

Individual suffering from SCD disorder has different level of crisis. The level of anemia crisis frequency has ranges from mild to moderate levels. In some cases they can develop severe anemia that are life threatening. Severe anemia in children can be caused by splenic sequestration of aplastic crisis.

1)Spleenic Sequestration Crisis

The spleen is one of the human body organs positioned at the upper side of the belly, and its functions are as follows: to filters germs in the blood, breaks up blood cells and develop or recreate new white blood cell. Splenic sequestration crisis occurs in children living with SCD disorder when the RBCs get stuck in the spleen thereby making a quick enlargement which brings about lesser circulation o blood to other cells and this result into anemia crisis. Austere pain may also occur in the left side of the belly to an enlarged big spleen which may cause palpation or wrong feelings by the patient [5].

2)Aplastic Crisis

This is also known as the fifth (5^{th}) disease. It is a mild rash illness caused by Parvovirus B19. It is also referred to as *erythema infectiosum*or *slapped cheek syndrome* because it is the fifth in the list of historical classifications of common skin rash illness in children. Aplastic crisis is common in children than adults. Parvovirus B19 is a very common children infection but in sickle cell disease patients; if fever, itching, running nose, headaches and swollen joints which are its numerous complications are not taken care of properly, it can cause delay or total halt of bone marrow development of new RBCs for a long period of time which results to severe anemia in children.

Severe anemia is not common to adults living with sickle cell disease disorder but they often experience other causes possibly related to severe anemic conditions such as breath shortage, exhibiting tiredness, dizziness or pale skin.

C.Infections

Human spleen is responsible for protecting the body from certain germs. In human's early life, sickle cells can damage, weaken or destroy the guiding function of the spleen. Patients with sickle cell disease disorder experiences or have damaged spleen at high risk of serious bacterial infections that can be life-threatening. These bacterial infections include but not limited to the following: pneumococcal disease (an infection caused by the Streptococcus pneumonia), hemophilus influenza (bacterial infection common to children under age 5, can be treated by *Hemophillus influenza type b – HiB*vaccine against meningitis), meningococcus (caused by *bacterium Neisseria meningitides* which carries high mortality rate in children if not treated), salmonella (a food poisoning bacteria), staphylococcus (common but usually found on skin, nose, hair and throats of even healthy

people), chlamydia (common sexually transmitted disease (STD) that can be easily cured if left untreated but can cause difficulty in women getting pregnant), mycoplasma pneumonia (a form of bacteria that causes infection of the respiratory system), etc. These bacterial infections can cause serious infections from blood infection (septicemia) to lung infection (pneumonia), infection of the covering of the brain and spinal cord (meningitis), bone infection (osteomyelitis). The latter is very common among Nigerians suffering from SCD disorder which has resulted to bone damages or severe bone ulcers leading to amputations [5].

D.Acute Chest Syndrome (ACS)

Sickling of the red blood cells or blood vessels can cause an individual suffering from sickle cell disease disorder to have shortage of oxygen to their lungs. When these occur, the tissues around the lungs get damaged and cannot exchange oxygen properly; and this is known as *Acute Chest Syndrome (ACS)*; where at least one segment of the lungs is damage. ACS frequently exhibits medical symptoms similar to pneumonia. In developed countries like US and UK, ACS has the greatest mortality rate in sickle cell disease patients after 2 years of age, and this is one of the high cause of paediatric intense care unit admissions in developed counters, and recently it was rated the second most collective cause of hospital admission after VOE [1], [5]. ACS is one of the most serious complications of SCD disorder as witnessed in developed countries and should be treated seriously with proper management and treatment in all the hematology departments of Nigerian hospitals. Patients with SCD disorder usually develop these conditions few days after a painful crisis starts. Lung infection may follow these conditions in some cases. There are various signs and symptoms associated with ACS which include painful and sharp chest pain, fever, breath shortage, cough, and in some cases rapid breathing.

E.Stroke

In Nigeria, this complication has been reported of late to be common in SCD patients. This complication is dangerous as it does not give any signs before attacking the SCD patient. Recent records of high morbidity and mortality has been reported with stroke [5], [6]. In some quarters, this has cause emotional breakdown to victims of SCD and their families and oftentimes the loss of life. In HbSS and HbSC patients in Nigeria, the occurrence of overt stroke is about 11% in ages less than 20 years and up to 30% are more frequent in silent cerebral infarct patients [5], [6]. A silent infarct (SI) is known as laceration on magnetic resonance imaging (MRI) consistent with unknown infarction but without focal neurologic deficit lasting longer than 24 hours. Silent infarcts are associated with cerebral weakness, decrease in intellectual abilities, low academic accomplishment, and high risk rate for subsequent infarction (death tissue due to lack of oxygen) [1], [5], [6].

Other complications of SCD disorder are depicted in Figure 4 which includes amongst others brain complications, eye problems, heart disease, pulmonary hypertensions, kidney problems, priapism (common in male - prolonged or painful erection), gallstones, liver complications, bone ulcers especially in legs (osteomyelitis – common in Nigeria), joint complications, hip joints crushing, delayed growth during puberty periods, pregnancy infections, and mental health (frustrations, depression and isolations).

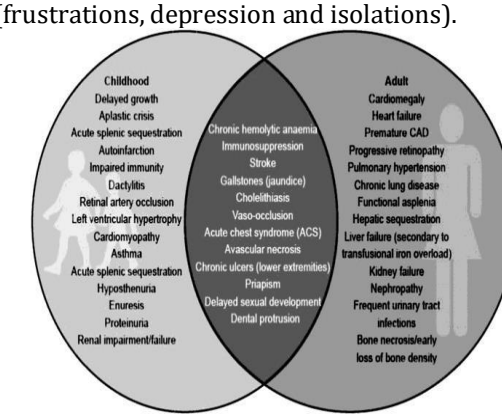

Fig. 4.Sicle Cell disease disorder Complications from Childhood to Adulthood [1] (Keys: coronary artery disease (CAD)

VI.RELATED WORKS

Several research works have addressed the study on SCD disorder especially in developed countries and few developing countries particularly in Africa. The most relevant and close related work to this paper is that of [16] where a thorough research review was carried out on management of SCD disorder from childhood to adulthood in the US in 2013. Their work was able to demonstrate that managing SCD disorder and having proper health information on the disease is paramount. They were able to explicate several gaps in managing the transitions of SCD. Recommendations were also given in their paper review on the next line of action on SCD disorder focusing on discovering cost-effective preventive and comprehensive care measures and programs for adults living with SCD disorder.

Another closely related work is that of [3] where it was stated clearly the process of improving SCD transitions care from childhood to adulthood

through health information technology. The survey was carried out also in the US during 2012-2014 through an experimental scan and private interviews with focus groups created to report the success of the development of health information technology (HIT) enabled tool to be used for SCD patients during care transitions. Although, the environmental scan was not a success during the period of the survey as it does not reveal an existing suitable transition tool for the SCD patients, but recorded a minor success where patients, parents, providers, and IT experts saw the potentials, possibilities and applications of creating a tool to meet emergency departments (EDs) health information needs to improve care transitions of these SCD patients.

Other works such as [7], [10] and [15] focused on mortality rate in SCD patients, recent treatment guidelines for managing SCD disorder and morbidity in SCD patients respectively. Thorough studies were carried out on SCD disorder in these papers. [12] was another related study comparing better hematological indices and lower consultation rate of Cameroonian patients' suffering from SCD disorder in a co-heritance between α-thalassemia and sickle cell anemia disease; and their results were positive.

Additional related work that was done in Africa is that of [2] where the focus is on children. Their survey and review was done on available African data on mortality rate in children associated with the most common form of SCD disorder in Africa which is caused by the homozygosis of the beta globin (β-globin) gene S mutation commonly known as *SS* disease. They were able to justify the wrong impression that the SCD condition is associated with the high rate of child mortality in Africa. They carried out their work, in the absence of low contemporary and reliable data, on two studies namely the cross-sectional population survey and cohort studies. They reported that early-life mortality rate was about 50-90% in children with SS disease in Africa and as such involving child existenece procedures and programmes could be of advantage in improving the health information management with respect to death rate among children living with SCD. They also supported the presence of government involvement in creating health investments for child's screening and adequate prophylaxis during their crisis. In same line of mind with this paper, their work also supported adequate and timely data to be available to determine the child mortality in Africa with respect to these interventions and their cost implications. In their conclusion, they recommended blood test samples of already collected specimens of children suffering from SCD disorder covering different age groups with respect to other infections such as HIV, malaria, malnutrition to determine their effect on SCD patients' mortality in African children.

Another work was that of [17] trying to make a comparison between survival of adult patients with SCD in the wealthy environment to the underprivileged zones. The survey was comprehensively carried out in King's College Hospital, London, UK. Their survey also considered on cohort groups subjecting the patients to Hydroxyurea therapy and some to blood transfusion. Their report shows that there are adequate and positive improvements and chances of survival for adult patients living with SCD. Their work was able to calculate mean numbers of survival of patients living with hemoglobin SS disease (HbSS) and other forms of SCD disorder (i.e. HbSC, HBSβ^+, HBSβ^0, and α-globin genotypes) in a wealthy environment in a space of 10 years (from 2004-2013) and it was confirmed that positive result were discovered which is an indication that adequate and proper health information management of SCD disorder can improve life expectancy of victims of the disorder even among adults. Their conclusion was a good landmark for adults living with SCD disorder which shows a life expectancy of 80 years for men and 84 years for women. This survey type is needed in Nigeria in order to get updated information on morbidity and mortality in SCD among adult patients.

Other related works are that of [5]–[11], [13], [14], [19], and [20] showing various epidemiology studies of SCD disorder. The various works were carried out at national and global levels and were able to demonstrate that there is no adequate or timely data estimates of population affected by either the heterozygote (AS) or the homozygous (SS) neonates. Some results were shown demonstrating evidence based estimates from different angles and scales using uncertainty measures.

VII.SCDMANAGEMENT IN NIGERIA

Comprehensive health information management of SCD patients through Information and Communication Technology

(ICT) in Nigeria is of great importance in this dispensation. Various management programs have been in place in the developed countries which has proven that ICT involvement in SCD disorder management have positive impacts on the SCD patients by reducing the complications as well as total annihilation of morbidity and mortality rate. Acute and longlasting pains are common

complications of sickle cell disease patients in developed world and now in Nigeria require individuals living with SCD to seek treatment and necessary information on management of the disorder. Nevertheless, most of these long-lasting pains are due to vaso-occlusion. According to [1] different diagnosis of SCD disorder using systematic approach for its treatment and management of SCD-related pain were discussed. For individuals above 18 years of age in Nigeria, there is scarcity of dedicated resources available especially in the rural areas where acute VOE needs urgent and aggressive treatment [5]. Some customary treatment such as opioids, non-steroidal anti-inflammatory drugs (NSAIDS) such as Pentazocine, diclofenac, etc., and proper hydration can be adequately employed to avert these complications. Table 2 shows a breakdown of both primary and secondary precautionary measures in young adults and adults with SCD disorder [1].

These programs illustrated in Table 2 can also be implemented in Nigeria and having the right health information management through ICT is supreme. The following are some of the tested and proven management programs for SCD disorder [1][5][6] [20]. They are:

- Folic Acid and Penicillin: Daily intake of folic acid is recommended for every child above 5 years old. In addition, in infants living with SCD disorder, daily penicillin intake, due to immature system of their body makes the system prone to early childhood illness, is also recommended.
- Inhibition of Malaria disease: In Nigeria, malaria attack is one of the prevalent causes of sickle cell anemia crisis in individuals with SCD but this does apply to those with sickle cell traits (HbAS). Therefore, it has been recommended for patients living with SCD, especially in Nigeria, to receive a chemoprophylaxis anti-malarial therapy through their lifetime.
- Vaso-occlusive crisis: This is common in patients with SCD disorder; however the rate of frequency, severity and durations varies from one individual to another. These VOEs are treated with proper pain reducing medications which require pain management with opioids administration at regular intervals during VOEs crisis until the pain subsided. Others can be observed under NSAIDs treatment (e.g. diclofenac, Pentazocine, etc.). In emergency departments (EDs) cases or hospital admissions, intravenous opioids such as patientscontrolled-analgesia (PCA) are recommended to be used. In other cases, Diphenhydramine is also recommended to prevent itching associated with the use of some opioids (e.g. the quinine vaccine family for malarial parasite treatment.
- ACS: As discussed earlier, ACS is much more similar to VOEs crisis and as such must be taken seriously and well managed. In such cases antibiotics (e.g. quinolone or macrolide) should be added to supplement for hypoxia (lack of oxygen) [16], [20].
- Hydroxyurea (HU): This is the first approved and established preventive pharmacology treatment for patients living with SCD disorder with persistent VOEs both in infant and adults. In a study carried out in 1995, HU has shown to reduce the frequency rate and severity of VOEs crisis, and also possible increase of survival time in a study carried out in 2003 [20]. The process was done by reactivating the fetal hemoglobin in place of hemoglobin S gene that triggers sickle cell anemia. HU has been previously used as a chemotherapy agent, however, it has been warned that long-term usebe harmful but from research its benefits outweigh the risks [20]. Hence, it is reasonable

Other management programs include blood transfusion, bone marrow transplant, and proper home management. In support for home management therapy, the following various home management programs for SCD patients are illustrated which include different steps not limited to controlling pain symptoms but also preventing other complications caused by the disease emanating [5]:

- Daily hygiene to children to guide against childhood infection.
- Regular does intake of antibiotic for SCD children up to age 5
- Children immunization on schedule e.g. Hemophilus influenza type B [Hib], hepatitis B, pneumococcal, and influenza vaccine should be given.
- Regular avoidance of dehydration to guide against sickling blood vessels.
- Every SCD disorder patients are encourage to cultivate water intake habit and other fluids before, during, and after exertion and when in the heat.
- Avoid all dehydration factors such as alcohol consumption
- Guide against conditions that reduce the oxygen levels in the blood.

- Guide against long stay in high altitudes e.g. an unpressurized airplane or high mountains Avoid cigarette smoking.
- Stress management is highly recommended through adequate sleep and thus reduces fatigue.
- Cold environment and temperature must be avoided e.g. long stay in air-conditioned room or car or long raining periods as this can triggers painful event.
- Proper education on the disease and regular eye examination is recommended
- Develop the practice to identify serious symptoms of the disease. This can be achieved by partnering with your medical doctors and hematologist. Serious warning indicators include fever higher than 101°F (38.33°C); austere cough, difficulty in breathing or shortness of breath; chest pain; severe abdominal (belly) pain; consistent vomiting or persistent diarrhea; sudden increase in the size infants spleen; increased or continuous paleness; lightheadedness; sudden weakness; unexpected numbness or tingling in the hands, feet, fingers, or toes; impulsive development of poor walking balance or coordination.
- Other indicators are confusion; garbled speech or inability to speak; impulsive changes in vision; longlasting headache; loss of consciousness; persistent erection of the penis (priapism) lasting more than 3 hours or extremely painful; severe pain that can't be relieved with the usual painkilling prescription drugs or other pain-relief methods.
- Consume proper diet and dietary supplements.
- Supplements such as folic acid as prescribed by medical doctors must be observed to aid the development of new red blood cells by the bone marrow.

TABLE II. PRIMARY AND SECONDARY PRECAUTIONARY MEASURES FOR SCD IN INFANTS AND ADULTS (ADAPTED FROM [1])

Precautionary Measures	Management procedure
Immunization – this is highly recommended for HbSS patients with damaged spleen	• Patients with HbSS disease are advised to be immunized yearly against influenza. Intake of pneumococcal vaccine every 5 years, and a meningococcal vaccine series (2-doses primary series controlled 2 months apart for persons under 2 years • Hepatitis A and B vaccines are recommended to patients that have undergone one blood transfusion of the other.
Sickle cell retinopathy screening	A routine check of this must be performed once a year
Hypertension screening	• This must also be performed at least once annually • Patients should be managed on antihypertensive treatment with the aim of lowering blood pressure to ≤140/90 mm Hg on an average
Proteinuria check (Urine assessment)	Patients' urine is expected to be checked at intervals of their checkups and in case of tenacious proteinuria, renal specialist is consulted. The use of ACE inhibitors can also be of help.
Pulmonary Hypertension screening	This is not common with SCD disorder carriers (HbAS asymptomatic) however, it is recommended for sickle cell disease patients (HbSS-symptomatic). Pulmonary hypertension poses a higher mortality risk in vaso-occlusive crisis periods in SCD patients.
Blood transfusion history (10–20 units of blood)	• SCD patients in these categories are at high overload risk • Iron overload measures by ferritin is not always accurate and also increases during long-lasting pains. A level of N1000 ng/mL indicate an overloaded state in SCD patients and patients susceptible to blood transfusion should avoid this while iron chelation should be administered

Ischemic stroke	• The many factors surrounding ischemic stroke in older adults with SCD are not adequately assumed as this has shown similar risk factors in non-SCD patients in this age group • In another perspective, part played by the acute and chronic transfusion in adults has not been adequately defined; therefore, treatment for stroke in adult patients with SCD is based on past documentation for paediatric patients with SCD. In addition, patients should undergo transfusion exchange for acute stroke through consultation with a hematologist • Reports have shown that patients living with SCD experiencing stroke may as well have cognitive deficits that cause difficulty in understanding discharge instructions and remembering to keep follow-up appointments. In some cases it leads to death if proper management is not administered

Keys: homozygous sickle hemoglobin mutation (HbSS), andotesin converting enzyme (ACE)

VIII.PROPOSEDMETHODLOGY

The involvement of information and communication technology (ICT) as a positive protagonist in health information management for SCD patients in Nigeria is highly needed for proper data management and health care implementation. This paper work is compiled to address the need to manage and interpret the various amounts of data that have been collated, yet to be collated as well as those generated by different genomic research in order to have a robust health information management through the use of ICT on SCD disorder in Nigeria.

Genes, cells, genetics and genomes are all interrelated terminologies necessary for carrying out this research work in providing a suitable database management for the SCD disorder in Nigeria. Genetics is the study of heredity which originated from Mendel's theory of inheritance of simple traits such as Cowpea colours. One of the major and central abstractions in genetics is the study of gene. Cells, as explained earlier in this paper, is an assessment of chemicals inside a sac bounded by a fatty layer called the *plasma membrane*. Genetics, a material in cells, is contained in a structure called *Chromosomes*. Genome is the hierarchical information that an organism passed to its offspring, represented in each of its cells. The representation is called DNA molecule (*Deoxyribonucleic acid*). DNA is a molecule that carries the genetic instructions used in the growth, development, functioning and reproduction of all living organism and many viruses. Each DNA molecule is a long chain of chemical instructions called *nucleotides* of four different types i.e. fA, C, T, Gg. The totality of all these information is referred to as *GENOME* [21].

In humans, the genomes consist of nucleotides which the molecular biology is given the task to:

- Extract the information contained in the genome of different organisms
- Elucidate the structure of the genome
- Apply this knowledge to the diagnosis and ultimately treatment of genomes disease
- Comparing the genomes of different species and explaining the process and mechanisms of its evolution

Therefore, the proposed methodology to support this paper work is to implement the use of bioinformatics techniques which is the process of using performance computing and mathematics technique to perform a thorough data management, research and analysis of large amount of DNA sequence data in order to find variables that affect the health, disease and drug response with respect to sickle cell disease disorder in Nigeria. The approach to be implemented is known as *Bioinformatics*.

Bioinformatics relates to biological molecules requiring knowledge in various fields. The threat of over simplifying a very complex issue like SCD disorder, the process of understanding genetic diseases typically profits through three stages

(a.)Recognition of the disease state or syndrome including an assessment of its hereditary character

(b.)Discovering and mapping of the related polymorphisms or mutations.

(c.)Elucidating the biochemical/biophysical mechanisms leading to the disease phenotype

In light of this, this paper would be standing on the methodology of using bioinformatics techniques, a computer-assistant method, to determine the combined effect of ICT on non-communicable disease like sickle cell disease in Nigeria. The proposed method would be implementing SQL – Structures Query Language and SAS (Statically Analysis System) software with MATLAB support

to carry out this work on health information management for SCD in Nigeria.

IX.RECOMMENDATIONS

This paper recommends prompt health information management (HIM) programs for SCD in Nigeria with emphasis in the transition stages. Adequate HIM programs through ICT should be employed in hospitals, clinics and rural health centres in Nigeria. Introduction of these HIM programs for appropriate awareness must be inculcate in tertiary institutions against patients' additions to some of the prophylaxis. Better understanding and management of the disorder will greatly reduce the high rate of morbidity and mortality in Nigeria.

X.CONCLUSION

The SCD disorder is one of the silent killer disease that grossly affects the country, as such, considerable vulnerabilities among patients and accumulating morbidities associate with transitions from childhood to adulthood challenges the proper management of the disease [1]. Management of this type of genetic disease requires proper counseling (adequate health information for individuals and families understand the risks and options), diagnosis (getting the right medical attention) and treatment (averting the morbidity and avoiding mortality of the disease).

In addition, SCD treatments in Nigeria are not adequately and seriously observed unlike other communicable diseases and the use of HU were underutilized compared to other developed countries. Therefore, adult patients with SCD disorder are expected to adequately have access to management services through ICT in various hospitals and clinics to forestall the high rate of morbidity and mortality in Nigeria. The use of health information management system can be the right solution.

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