Prevention and Management of Sickle Cell Anemia: A Medico-Social Perspective

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Certification

This is to certify that Omolara Oluwafunto Adeniyi with matric number LCU/PG/002669 carried out the discussion of this essay titled ―Prevention and Management of Sickle Cell Anemia: A Medico-Social Perspectice‖ in the Department of Social Work, Faculty of Management and Social Sciences, Lead City University, Ibadan, Oyo State for the Award of Master of Social Work (MSW).

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Dedication

I dedicate this project to the glory of God Almighty for his infinite mercy and love for me since the beginning and to the completion of my study.

Acknowledgement

Thousands of tongue cannot be enough for me to appreciate God almighty for all He has done on my behalf and for seeing me through this essay writing project on: ―Prevention and Management of Sickle Cell Anemia: A Medico Social Perspectice‖.

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ABSTRACT

Sickle Cell Disease is an age-old disease which has been known for hundreds of years in Africa. It was known in Africa by onomatopoeic names denoting the recurrent, unrelenting, and painful nature of the disease. Although, the symptoms of the disease could be traced to 1670 in a Ghanaian family, disorders of hemoglobin synthesis were unrecognized by the scientific community until 1910.

Sickle cell disease is the commonest single gene disease in Africa. It is common throughout the sub-saharan Africa, affecting up to 30% of people in some parts of the continent. Morbidity and mortality from this disease has remained unacceptably high in Africa whereas there has been a marked reduction in the burden of this disease in the developed countries. This reduction was not achieved through the use of sophisticated care such as bone marrow transplant, but through the adoption of transition programs, premarital counseling, public education, genetic education and counseling, and a Comprehensive Health Care Management protocol for sickle cell disease. This protocol of care emphasizes prevention of crises through effective management of the disease.

This discourse examines the dimension of SCD, tracing its genesis and implications, its psychological impacts, national burden and the way forward. The paper also examines various strategies for preventing SCD as well as mechanisms for coping or SCD management mechanisms. As this discourse is from the perspective of Master of Social Work, the paper sheds light on the social work profession and its role in the prevention and management of SCD.

Keywords: Prevention, Management, Sickle Cell Anemia

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List of Acronyms

SCD – Sickle Cell Disease

SCA – Sickle Cell Anaemia

RBC – Red Blood Cells

Hb – Hemoglobin

HBB – Beta-globin gene

ISCs – Irreversibly Sickled Cells

BPC – Bone Pain Crisis

NSAIDS – Non-Steroidal Anti-Inflammatory Drugs

TENS – Transcutaneous Electrical Nerve Stimulation

MOFS – Multi-Organ Failure System CVD –

Cerebrovascular Disease

CBT – Chronic Blood Transfusion

ACS – Acute Chest Syndrome

CHWs – Community Health Workers

WHO – World Health Organization

SW – Social Worker

Chapter One

Introduction

Sickle cell disease (SCD) is one of the most common genetic diseases worldwide and its highest prevalence occurs in Middle East, Mediterranean regions, Southeast Asia, and sub-Saharan Africa especially Nigeria [1, 2].

SCD is a chronic haemolytic disorder that is marked by tendency of haemoglobin molecules within red cells to polymerise and deform the red cell into a sickle (or crescent) shape resulting in characteristic vasoocclusive events and accelerated haemolysis. It is inherited in an autosomal recessive fashion either in the homozygous state or double heterozygous state. When inherited in the homozygous state, it is termed Sickle Cell Anaemia (SCA). Other known SCD genotypes include haemoglobin SC disease, sickle beta plus thalassaemia, and sickle beta zero thalassemia (which has similar severity with sickle cell anaemia), haemoglobin SD Punjab disease, haemoglobin SO Arab disease, and others.

In Nigeria, SCD forms a small part of the clinical practice of most general duty doctors, as there is gross absence of dedicated sickle cell centres. Thus, it may be difficult to keep abreast of current knowledge and practices in the treatment of SCD. The purpose of this essay therefore is to provide a comprehensive and concise review of SCD, prevention and its management from a medico social perspective and for physician education in Nigeria. Particular attention is given to its local epidemiology, clinical phenotypes and complications, current treatment guidelines, practice challenges, and recommendations for improved care.

1.1 Objectives of the Essay

The specific objective for this discourse is to examine the prevention and management of Sickle Cell Anemia from a medico social perspective. Hence, the specific objectives for this discourse are to:

1. Have an overview of Sickle Cell Disease, brief history and its genetic origin.
2. Examine the complications in Sickle Cell Anemia
3. Examine the chronic morbidities in Sickle Cell Anemia
4. Examine the concepts of Social Works and its functions
5. Examine the current challenges in Nigeria and how to control the sickle cell disease.
6. Examine the Psychological Impacts of the Sickle Cell Anemia on both the Patients, Caregivers and Family.
7. Examine the National Burden of Sickle Cell Disorder and the way forward
8. Examine the principles of Genetic Cousnseling
9. Examine the prevention and management measure of Sickle Cell Anemia
10. Evaluate the Impact of Community Health Workers on Sickle Cell Disease
11. Examine The Role of Social Welfare Counselling in the Eradication of Sickle-Cell Disease in Nigeria

1.2.An Overview of Sickle Cell Disease

Blood is a specialized body fluid and has four main components: plasma, red blood cells, white blood cells, and platelets. Hemoglobin (Hb) is a protein based molecule found in the red blood cells (RBC) that carries oxygen in our body and gives blood its red color. Normal red blood cells are biconcave, have no nucleus, and being flexible can easily change shape, which helps them to fit and move easily through the smallest blood vessels called capillaries. Sickle Cell Disease is an inherited red blood cell disorder, wherein the mutation in the gene

causes the formation of atypical hemoglobin. This causes RBCs to lose their normal shape and become C-shaped, like sickles or crescent moons and lose flexibility. These rigid, sticky cells can get stuck in small blood vessels and cause clogging of blood vessels causing slowing or blocking blood flow and oxygen to parts of the body. It is one of the most common monogenic disorders globally with an autosomal recessive inheritance. James Herrick first described the characteristic sickle-shaped red cells[3] and Linus Pauling and his colleagues[12] showed that sickle hemoglobin (HbS) had an altered electrophoretic mobility and defined the molecular disease in 1949. Normal red blood cells can live up to 120 days but sickle cells only live for about 10 to 20 days. The primary pathophysiology is based on the polymerization of deoxy HbS with the formation of long fibers within the RBCs causing a distorted sickle shape and leading to increased hemolysis and vasoocclusion by sickle red cells. Also, sickle cells get destroyed by the spleen because of their shape and stiffness. Sickled cells get stuck in this filter and die. With fewer healthy red blood cells circulating in the body, a person becomes chronically anaemic and sickled cells also damage the spleen. It results in various complications like anaemia (Sickle Cell Anaemia), frequent infections, pain, and swelling as well as chronic damage to various organs in the body including the brain, liver, lungs, etc.

Sickle cell disease is an age-old disease which has been known for hundreds of years in Africa. It was known in Africa by onomatopoeic names denoting the recurrent, unrelenting, and painful nature of the disease.[4] Translated, some of these names mean a child destined to die young or a child who brings sadness and pain to his parents.[5] Since a heterozygous mating produces a 25% chance of bringing forth a child with the disease during each pregnancy, a couple could have a series of pregnancies resulting in babies with the disease. In some ethnic groups, elaborate ceremonies were performed to prevent future reincarnations of

the child. It is still a feared disease in parts of Africa, although there is a greater understanding of the genetics and disease etiology.

Though once considered a disease of childhood with a mortality approaching 20% by age 3, diagnosis soon after birth and advances in clinical research have placed current life expectancies, on average, at more than 48 years.[6,7] Sickle cell disease is an inherited disease in which defective sickle-shaped red cells fail to carry adequate oxygen to tissues in the body. As a result of the sickling, the cells tend to block and damage the smallest blood vessels in the body, resulting in damage to organs served by those vessels.

Today, the prognosis is not as bleak for the patient with sickle cell disease as it was in the past. Various treatment options are now available to help the patient with this chronic disease cope with its many complicating aspects.

Although the symptoms of the disease could be traced to 1670 in a Ghanaian family, disorders of hemoglobin synthesis were unrecognized by the scientific community until 1910[8]. The first article to describe the sickle cell phenomenon appeared in 1910, when Herrick[9], a Chicago cardiologist, wrote a case report titled ―Peculiar elongation and sickle shaped corpuscles in a case of severe anemia.‖

The article contained a description of the symptoms and pictures of blood obtained from a 20-year-old patient from the West Indies. In 1927, Hahn and Gillespie[10] delineated the conditions affecting sickle cell formation in vitro, including pH, temperature, fixatives, tonicity, and others, and determined that loss of oxygen was responsible for the sickle shapes observed in the blood. They postulated that similar effects of hypoxia could occur in vivo, leading to cellular distortion with consequent hemolysis. In 1940, Sherman noted a birefringence of deoxygenated red cells suggesting that low oxygen altered the structure of the hemoglobin (Hb) molecule[11]. In 1948, Watson suggested that the scantiness of sickle

cells in the peripheral blood of newborns was due to the presence of fetal hemoglobin in the red cells[5]. In 1949, Pauling and colleagues[12] showed that hemoglobin from sickled cells had an abnormal mobility in an electric field. In 1949, Neel published a report establishing that sickle cell trait was the heterozygous state, and sickle cell anemia was the homozygous state for the same gene.[13] In 1956, Ingram[14] revealed that substitution of valine for the glutamic acid in the sixth position of the beta globulin molecule was responsible for the abnormal function of the molecule after deoxygenation. The hemoglobin S (HbS) molecule is a protein whose quaternary structure is a tetramer consisting of 2 normal α-globulin chains and 2 abnormal β-globulin chains. The pathology that leads to the sickle shapes of the red blood cells involves this molecule. After deoxygenation of HbS molecules, polymers of HbS form through hydrophobic interactions between the β-6 valine of a tetramer and the β-85 phenylalanine and β-88 leucine of an adjacent tetramer.[15] The HbS molecules aggregate upon deoxygenation to form polymer nuclei that become seeds for further polymerization. The polymerization of the sickle red cells takes place as the cells traverse the microvasculature [16]

Factors that increase the intracellular concentration of hemoglobin, factors that increase time spent in the microvasculature, and deoxygenation of the hemoglobin all contribute to increased polymerization. Increased levels of non-S hemoglobin such as HbF and HbA[5] slow the rate of polymerization and reduce the intracellular polymer content at any oxygen saturation.[16]

Epidemiological Considerations

About 5–7% of the global population carries an abnormal haemoglobin gene [17, 18]. The most predominant form of haemoglobinopathy worldwide is sickle cell disease. The greatest burden of the disease lies in sub-Saharan Africa and Asia [19].

The sickle cell disease was first recognized among persons of western African ancestry. The sickle cell trait is seen in 10% to 30% of people in equatorial Africa but infrequent in northern and southern Africa [12]. The HbS gene is distributed worldwide, occurring around the Mediterranean in Sicily and other parts of southern Italy, northern Greece, Turkey along the south-east coast, the north African coast, in Saudi Arabia especially the eastern province, Iran, and throughout central India.[2]

Approximately 1 in every 400 to 500 African Americans has sickle cell disease. [20] An estimated 80,000 Americans have the disease and about 9% of African Americans have the trait. One in every 1,000 to 14,000 American Hispanic children are born with sickle cell disease.[21] In India, the sickle cell gene is present in all tribal populations who inhabit hilly forest areas where falciparum malaria is common.[22] It presents as a lethal disease in Africa but benign or mild in India and Saudi Arabia.[23,24] It is the most prevalent inherited monogenic pathology in South America and it is estimated that 2% of the population of Brazil and 6% to 9% of Brazilians of African descent are heterozygous for the HbS gene, with 700 to 1,000 new cases yearly [25]. The HbS gene is found at a frequency of 20% to 30% people in some villages in northern Greece, 25% in the Qatif oasis of eastern Saudi Arabia, and in 20% to 30% of many communities in the Indian states of Orissa, Madhya Pradesh, and Maharastra.

The prevalence of sickle cell trait ranges between 10 and 45% in various parts of sub-Saharan Africa [26–28]. In Nigeria, carrier prevalence is about 20 to 30% [29, 30]. SCD affects about 2 to 3% of the Nigerian population of more than 160 million [29]. Recent estimate from a large retrospective study by Nwogoh *et al*. in Benin City, South-South Nigeria revealed an SCD prevalence of 2.39% and a carrier rate of about 23% [31].

1.3. Brief History and Genetic Origin of SCD

In 1874, Dr. Horton, a Sierra Leonian medical Doctor, reportedly gave the first description of clinical symptoms and signs which is now referred to as sickle cell disease[32]. Herrick, a Chicago physician, also gave a formal description of the disease in 1910 when he observed abnormal sickle shaped red cells in the blood of a dental student from West Indies who had anaemia[9]. In 1927, Hahn and Gillespie observed that sickling of red cells was associated with conditions of low oxygen tension. In 1949, Linus Pauling and colleagues demonstrated that haemoglobin in these patients was different from normal subjects using protein electrophoresis [33].

However, Venon Ingram and J. A. Hunt in 1956 sequenced the sickle haemoglobin molecule and showed that the abnormality was due to valine substitution for glutamate on the 6th position of the sickle beta-haemoglobin gene. Marotta and coworkers in 1977 showed that the corresponding change in codon 6 of the beta-globin gene was GAG to GTG [33]. Since then, further insights have been gained into understanding the origin, complex pathophysiology, and treatment of the disease through molecular biology techniques.

Africa and Asia are considered as the birthplace of the sickle cell mutation. Sickle cell disease is believed to be a consequence of natural mutation of the beta-globin gene (HBB) affecting the gametes and transferred to subsequent generations. Using restriction fragment length polymorphism analysis, four main African haplotypes and one Asian haplotype of the beta-globin chain genes have been characterized and are believed to originate differently in these regions. The main African haplotypes include Senegal, Benin, Bantu (central-African republic), and Cameroon haplotype [34–37]. The Bantu haplotype is associated with the most severe disease phenotype while the Asian (also called Arab-Indian) haplotype is associated with a mild phenotype [38].

SCD is found in other parts of the world including USA and Europe due to migration and interracial marriages [19, 39]. The high prevalence of SCD in sub-Saharan Africa has been attributed to survival advantage conferred by the sickle cell trait against *Plasmodium falciparum*. Resistance of individuals with sickle cell trait to *Plasmodium falciparum* creates a selective pressure that has maintained the sickle cell gene within human populations in malaria endemic regions like sub-Saharan Africa. This phenomenon is termed balanced polymorphism

[40, 41].

1.4 Aetiopathogenesis of Sickle Cell Disease

SCD is a qualitative haemoglobinopathy resulting from a structural change in the sequence of amino acids on the beta globin chain of the haemoglobin molecule due to a point mutation. The sickling mutation causes a single base change from adenine to thymine on the 17th nucleotide of the beta globin chain gene (HBB). This invariably translates into substitution of valine for glutamate on the 6th amino acid of the beta globin chain. The abnormal biochemistry of this mutant haemoglobin induces polymerization of Hb S molecules within the red cells, so called sickling. On the sickle haemoglobin, the glutamate protein molecule, which is hydrophilic, polar, and negatively charged, is replaced by a less polar, hydrophobic, neutral amino acid, valine. Under deoxy conditions, the abnormal valine residue causes intraerythrocytic hydrophobic interaction of sickle haemoglobin tetramers, leading to their precipitation and polymer formation, so called gelation [42]. Eventually, all cytosolic haemoglobin molecules precipitate into seven (one inner and six outer) double strands with cross-links which are called tactoids. Upon reoxygenation, unsickling occurs and the red cell assumes its normal shape. However, repeated sickling and unsickling of the red cell damages the red cell membrane, due to herniation of sickle haemoglobin polymers through the cytoskeleton, thus rendering the red cell permanently sickled. These appear as irreversibly

sickled cells (ISCs) on peripheral blood cytology. The kinetics of red cell sickling is highly heterogenous.

Several variables are known to affect the rate and degree of sickling of the red cells. Intracellular dehydration of sickle red cells increases mean cell haemoglobin concentration (MCHC) [33]. Higher MCHC favours sickling. As such, very high Hb S level of about 80 to 90% seen in the homozygous disease is associated with a worse disease while the presence of alpha thalassemia (one or two gene deletions) ameliorates the disease. Another variable is the presence of other interacting non-sickle haemoglobin. Of note is fetal haemoglobin (Hb F). Higher proportion of Hb F is associated with mild disease. When present, high levels of Hb F are uniformly dispersed within the red cell and it retards the sickling process. Thus, coinheritance of sickle haemoglobin with hereditary persistence of fetal haemoglobin (HPFH) is associated with mild disease[43]. Similarly, this advantage is positively utilized through clinical use of fetal haemoglobin inducing drug such as hydroxyurea. Vascular beds that have intrinsically sluggish venous outflow such as bone marrow, spleen, or inflamed tissues are at higher risk of infarctive events due to prolonged microvascular transit time [44].

Whenever and wherever microvascular transit time becomes longer than sickling delay time, sickling and vascular occlusion become imminent. Intracellular pH is another important variable. With acidosis, the haemoglobin molecules give off their oxygen more readily and sickling occurs more readily.

Repeated sickling of the red cell induces cellular injury which has been shown to activate membrane ion channels such as the Gardos pathway (calcium gated potassium channels) and KCL cotransporter [44]. There are influx of calcium ions and efflux of potassium and water, hence intracellular dehydration. High intracellular calcium levels provoke activity of proteolytic enzymes such as phospholipases and proteases causing the digestion of membrane

phospholipids and proteins, respectively. Subsequently, there is perturbation of the membrane lipids with exteriorization of lipids such as phosphatidyl serine and ethanolamine which are normally located in the inner leaflets of the membrane lipid bilayer [45].

The diverse clinical heterogeneity of SCD is related to two main pathogenetic processes: chronic haemolysis and high viscosity/vascular occlusion. Infarctive events in SCD result from erythrostasis caused by rigid sickled cells in various vascular beds especially organs with sluggish blood flow such as the spleen and the bone marrow. Capillaries are about 2-3 microns in diameter. Sickle cells due to loss of flexibility are unable to transit the microvasculature, hence vessel occlusion. Aside from these mechanistic processes, sickle cells are also shown to exhibit increased adhesiveness to vascular endothelium, leucocytes, platelets, and themselves [46–48]. Sickle reticulocytes are even more adhesive to the endothelium than sickle discocytes [49]. Molecular interactions between the red cells and the vascular endothelium include CD36 and thrombospondin, VLA4 and VCAM-1, respectively [49, 50]. Fibronectin and von Willebrand factor are also involved in these interactions. Currently, it is known that increased adhesiveness of different cellular surfaces with formation of hetero cellular aggregates is believed to propagate the phenomenon of vascular occlusion, especially in the postcapillary venules [45]. Also note that destruction of red cellmembrane causes exposure ofmembrane proteins, thereby inciting autoantibody formation. These antibodies such as IgG anti-band 3 antibodies are believed to promote erythrophagocytosis[44].

Episodic microvascular occlusion in sickle cell disease even in steady state results in ischaemic-reperfusion injury which sets the stage for an increased inflammatory tone, thus significant elevations in total leucocyte counts, platelet counts, and positive serum acute phase reactants. Even in steady state, SCD is a chronic inflammatory condition; the attendant inflammation induced oxidative stress further contributes to progressive tissue damage[51, 52].

The leucocytes in SCD also express higher levels of L-selectins and also have stimulated adhesiveness. Increased adhesiveness coupled with phosphatidyl serine exposure on the red cell surface makes SCD a procoagulant and a hypercoagulable state [53].

Erythrocyte lifespan in SCD averages about 16–20 days in contrast to about 100–120 days in normal state[54, 55]. Haemolysis in sickle cell disease is both extravascular and intravascular. Abnormal shape of the ISCs creates an abnormal rheology, associated with heightened clearance by the reticuloendothelial system. Because of their increased fragility and reduced deformability, some red cells undergo intravascular haemolysis. High plasma haemoglobin level is associated with low haptoglobin levels and high levels of lactate dehydrogenase (LDH), arginase-1, and AST [56]. Plasma haemoglobin is an avid scavenger of nitric oxide (NO). High plasma haemoglobin levels resulting from chronic haemolysis reduce NO bioavailability. Normally, NO relaxes the endothelium and maintains vascular tone (vasodilator) [56].

Low circulating levels of NO propagate vasospasm which is observed even in large vessels in SCD. Contributing to this is dysfunction of endothelial nitric oxide synthetase [56]. This is the underlying basis of vasculopathic complications such as cerebrovascular diseases, priapism, pulmonary hypertension, and chronic leg ulcers. Arginase 1 is normally involved in formation of urea in protein excretion. Accelerated haemolysis of red cells leads to higher levels of arginase 1. As such, more ornithine is produced, further depleting plasma arginine levels.

Excess ornithine is channeled to alternate pathways which produce excess prolenes and polyamines. These byproducts promote endothelial smooth muscle proliferations, further narrowing the vascular chamber[57]. Chronic haemolysis results in excess breakdown of

haemoglobin molecules and high levels of bilirubin, which is associated with formation of bilirubin pigment stones in the gall bladder (cholelithiasis).

Some authorities have attempted to categorize SCD patients into two clinical subphenotypes based on the overriding pathogenic process [44, 58]. Clinically, there is some degree of overlap between the two groups. Some patients experience more of viscosity/vasoocclusive complications and tend to have higher baseline haematocrit levels. Others experience more of vasculopathic complications due to more intense haemolysis associated with a lower baseline haematocrit [33, 58].

1.5. Clinical Phenotypes and Complications in Sickle Cell Disease

There is marked intra-individual and inter-individual variability in SCD. Clinical heterogeneity of the disease has been explained by both genetic and environmental factors. Known genetic factors contributing to variations in clinical severity of the disease include the pattern of sickle cell inheritance, nature of b-globin haplotype, Hb F level, and FCP loci [34].

Other modulators of the disease include presence of alpha thalassemia and other probable genetic influences as well as environmental factors such as access to optimal health care, ambient living conditions, and availability of finance [34].

Physical Effects of Sickle Cell Disease. Body habitus in SCD ranges from a normal build to a tall, lanky physique depending on the clinical severity. Other physical changes include prognathism, arachnodactyly, and increased APchest diameter (barrel chest) [59, 60]. In childhood, sickle cell patients may be shorter or smaller than normal. Puberty is often delayed but considerable growth takes place in late adolescence such that adults with sickle cell anaemia are at least as tall as normal [33]. However, adults that have suffered vertebral infarction and collapse may be shorter than normal. Many of these physical changes are due

to the chronic hypoxaemia associated with severe anaemia. Severe haemolysis in infancy causes marrow hyperplasia of the skull and facial bones, resulting in frontal bossing, prognathism, or malocclusion[61]. The abnormal facies results from extension of the marrow into the cortical bone causing widening of the diploe spaces and thinning of the bone cortex. The chronic haemolytic process is associated with pallor, jaundice, splenomegaly in early childhood.

Acute Sickle Syndromes/Complications

Bone Pain Crisis (BPC). BPC is the most consistent and characteristic feature of SCD [62]. The pain results from activation of nociceptive afferent nerve endings in the ischemic bones. Commonly affected bones include the long bones such as femur and humerus, vertebrae, pelvis, ribs, and sternum [62]. Multiple sites may be involved. An early manifestation of bone infarction is the hand and foot syndrome. This is characterized by dactylitis involving the small bones of the hand or foot, marked by diffuse swelling over the involved area. It often resolves spontaneously within one to two weeks and is rare after 2-3 years of life. Frequency of bone pain crisis is higher in patients with homozygous sickle cell disease, low Hb F, and higher baseline haemoglobin. It is said to be more common in young adults but its frequency tends to wane at older ages. Pain episodes vary in intensity and tend to resolve within a few days. In about 57% of cases, no precipitating factor is identified [63]. However, known precipitants include exposure to cold, dehydration, intercurrent infections such as malaria, physical exertion, tobacco smoke, alcohol use, hard drugs, high altitude, hypoxic conditions, physical pain, pregnancy, hot weather, emotional stress, or onset of menses [70, 64]. Suggested treatment guideline for uncomplicated BPC is presented as follows.

Treatment Guidelines for Bone Pain Crisis

i. Principles of treatment include adequate analgesia, hydration, warmth, prophylactic or therapeutic antibiotics if pyrexial after necessary culture samples are taken, as well as oxygenation if hypoxic (SpO2<90%) [64–67].

ii. Oral hydration must be adequate with at least 1.5 L/m2 of water based fluid per day in children and 60–70 mL/kg in adults. If parenteral, not more than 1.5 times maintenance is given in order to prevent volume overload considering baseline anaemia in most patients [68, 69].

iii. Patients and parents should be encouraged to keep a stock of simple analgesics at home in event of a painful episode. However, mild to moderate pain that does not succumb to home-based oral analgesia and hydration within 2 days requires hospitalization.

iv. Analgesia should be commenced within 15 to 30 minutes of presentation in the emergency room or day hospital. Effective analgesia should be achieved within 1 hour. There should be an ongoing assessment of analgesic efficacy every 30 minutes until pain is controlled, thereafter every 2 hours [68, 69].

v. Treatment should be individualized. The choice of analgesia depends on the severity of the pain and the patient‘s prior analgesic needs/history.

vi. Nonopioids such as simple paracetamol and NSAIDS (non-steroidal anti-inflammatory drugs) may be used in mild VOC. Weak opioids such as tramadol and DF118 (dihydrocodeine) are used for moderate pain while severe pain requires stronger opioids/narcotics such as morphine [64, 65].

vii. Adjuvants for pain control help in achieving better analgesia. They may include mild sedatives such as promethazine or diazepam. Combination of paracetamol or NSAIDS with opiates gives better analgesia because of their synergistic actions. Oversedation

should be avoided. Laxatives should be prescribed for prevention and treatment of constipation, a side effect of opioid use.

viii. More than five to seven days of sequential NSAID use should be avoided to reduce the risk of peptic ulceration and GIT haemorrhage. Also, NSAIDS are potentially nephrotoxic and are better avoided in established renal disease.

ix. Severe VOC requires parenteral opioid analgesia and hydration in a hospital setting. The dose of the analgesia should be titrated with the severity of the pain until adequate control is achieved in a fixed dose schedule (FDS), interspersed with short-acting agents for breakthrough pains.

x. Prophylactic incentive spirometry is recommended for prevention of acute chest syndrome especially in BPC involving the chest wall. In the absence of a spirometer, 10 deep breaths every 2 hours while awake between 8 a.m. and 10 p.m. are an alternative.

xi. If pain persists, patient controlled analgesia (PCA) should be considered where available. PCA is sparsely available in Nigeria, except in very few private facilities. PCA reduces the risk of pain under-treatment.

xii. Short-acting opioids in clinical use include tramadol, morphine, hydrocodeine, hydromorphine, fentanyl, oxymorphine, and oxycodeine. Longer acting opioids include methadone and slow release preparations of tramadol, morphine, and oxycodeine. Access to a wide range of opioids may not always be readily available in Nigeria; however, the available ones should be used.

xiii. In difficult cases, where pain is unremitting after 48 hours of well conducted analgesia, exchange blood transfusion (EBT) may be offered [70].

xiv. Hospitalization for severe BPC occurring on 3 or more occasions per year is an indication for initiation of hydroxyurea therapy or chronic transfusion therapy in patients that are intolerant of hydroxyurea.

xv. Pain in SCD is majorly nociceptive in origin. However, it may also have a neuropathic component marked by tingling/burning sensation or numbness. In such cases, drugs such as pregabalin or carbamazepine will be useful[65].

xvi. Pain control in SCD is essentially pharmacologic.

However, nonpharmacologic measures such as physical therapy with heat or ice packs, relaxation, distraction, music, menthol rub, meditation, and transcutaneous electrical nerve stimulation (TENS) are also helpful [65].

Treatment of acute sickle cell pain in a dedicated day hospital is associated with better outcome due to prompt triage and familiarity with analgesic needs of individual patients, hence reduced risk of pain under-treatment. Invariably, there is reduction in overall patient admission rates and better outcomes compared to emergency room settings[71–73].

However, most institutions in Nigeria lack daycare settings for management of sickle cell crisis. Furthermore, it is important to note that BPC may be complicated by a concurrent hyperhaemolytic crisis with resultant acute severe anaemia or may even progress to acute chest syndrome or multiorgan failure syndrome (MOFS).

MOFS is defined as sudden onset, severe organ dysfunction simultaneously involving at least two major organ systems (such as the liver, lung, and kidney) in the setting of an acute sickle cell crisis. MOFS is partly explained by significant vasoocclusive events in vital organs with major functional compromise and organ failure[74]. This life-threatening complication requires immediate intensive care and a multi-specialist attention including the intensivists, nephrologist, hepatologist, respiratory physicians, and others.

Acute Abdominal Pain. Acute abdominal pain in SCD may be due to sequestration crisis, vasoocclusion of mesenteric vessels, gall bladder/biliary tract disorders, or other non-SCD specific causes. In a Nigerian study by Akingbola et al., the aetiology of acute abdominal pain in a population of adult Nigerian SCD patients presenting in a tertiary facility were found to include SCD related complications such as abdominal VOC and acute cholecystitis, as well as other infective causes such as cystitis, gastroenteritis, appendicitis, and bowel obstruction[75]. About 38% of the cases were due to abdominal infarction/crisis [75]. In Ile-Ife, Akinola et al. observed abdominal pain due to VOC in 26% of sickle cell anaemia patients [76]. Microvascular occlusion may involve the mesenteric bed causing ischaemic abdominal pain. Abdominal pain of presumed vasoocclusive origin is termed abdominal crisis. Girdle syndrome, otherwise called mesenteric syndrome, is a rare complication owing to extensive collateral blood supplies to the mesentery and bowel wall [77]. Girdle syndrome or mesenteric syndrome is said to be present when there is an established paralytic ileus, which may be associated with vomiting, silent distended abdomen, dilated bowel loops, and air-fluid levels on abdominal radiography. Typically, a patient with girdle syndrome presents with generalized abdominal pain and rarely in shock if there is massive bowel gangrene (third space losses).

Other abdominal findings may include localized or rebound tenderness, board-like rigidity, and lack of movement on respiration. Abdominal radiography and ultrasound scan are helpful as well as investigations to rule out differentials such as pancreatitis, acute appendicitis, cholecystitis, biliary colic, splenic abscess, ischaemic colitis, and other forms of acute abdomen. Intravenous hydration, analgesia, and antibiotics are indicated. For an established ileus, NPO (nothing by mouth) should be commenced, as well as nasogastric aspiration, if there is vomiting. Urgent surgical opinion should be sought. Exchange blood transfusion may be necessary in mesenteric (girdle) syndrome. Biliary/gall bladder anomalies commonly

observed in SCD include cholelithiasis and biliary sludge [78]. Recent studies among Nigerian patients observed an age-related prevalence of about 5 to 10%for cholelithiasis [79, 80].

Visceral Sequestration Crisis. Infants and children less than 7 years are at greatest risk of sequestration crisis especially splenic sequestration. Children above this age group and adults are at less risk of splenic sequestration because the spleen tends to become fibrotic with repeated infarctions and cannot enlarge[43]. However, in haemoglobin SC disease, older children and adults can experience sequestration crisis. If not corrected rapidly, acute sequestration results in hypovolaemia, severe anaemia, and possibly death. A typical patient is irritable with rapidly enlarging spleen or liver and pain in the upper abdomen. Features of acute anaemia include worsening pallor, generalized weakness, and tachycardia. Early presentation in the hospital and close monitoring are important. Blood transfusion is necessary if haemoglobin level falls 2 g/dL below steady state haemoglobin levels or evidence of cardiac decompensation.

A major sequestration crisis is defined by haemoglobin level below 6 g/dL. There is risk for recurrence; therefore parents and caregivers must be taught how to examine the child‘s spleen regularly and report any abnormal finding to the physician immediately. Splenectomy is recommended after the second episode in children above two years of age. For children below 2 years of age, chronic blood transfusion should be offered [81]. Majority of SCD sequestration crisis involves the spleen. Though less common, the liver and lymph nodes may also be sites of sequestration in SCD.

Aplastic Crisis. Aplastic crisis usually occurs in those less than 16 years of age. It is commonly caused by parvovirus B19 infection which causes transient selective suppression of erythroid progenitors. In Nigeria, prevalence of parvovirus B19 infection is shown to be similar among sickle cell patients and the general population [82]. In normal subjects,

parvovirus B19 infection is asymptomatic. However, in patients with chronic haemolytic anaemia such as SCD, parvoviral infection is potentially devastating. Reticulocytopenia often lasts about 7 to 10 days, followed by spontaneous remission with reticulocytosis. Aplastic crisis may follow a recent upper respiratory tract infection and the patient may have flu-like symptoms such as headache, mild fever, and lethargy. Other findings include pallor and worsening anaemia. The condition is self-limiting. Treatment is to give red cell transfusion support until erythroid activity resumes.

Worsening (Acute) Anaemia in SCD. Baseline haemoglobin level in sickle cell disease ranges between 6 and 9 g/dL [83]. A study among adult SCD patients in Lagos by Akinbami et al. shows mean steady state haemoglobin levels of 7.92 •} 1.49 g/dL [84]. Average haemoglobin concentration of an SCD patient over a minimum of 4 weeks is considered steady state (or stable) haemoglobin level in the absence of any form of crisis in the preceding three months. Anaemia in SCD is usually well compensated. Occasionally, some patients have high steady state haematocrit level above 10 g/dL and they tend to present with more vasoocclusive complications. This has been termed high haematocrit syndrome [45].

Therapeutic phlebotomy when haemoglobin level is higher than 12 g/dL may benefit such patients[78]. However, in most other patients, significant decline of more than 2 g/dL below steady state level has functional consequences. Untreated severe anaemia is symptomatic and may precipitate heart failure. Causes of worsening anaemia in sickle cell disease may include hyperhaemolysis from any cause, aplastic crisis, megaloblastic crisis, iron deficiency, haemorrhage, renal failure, sequestration crisis, and extreme bone marrow necrosis. Treatment requires both definitive and supportive care. If patient shows signs of cardiac decompensation, blood transfusion should be given. The cause of worsening anaemia should be sought and treated accordingly.

Cerebrovascular Disease (CVD). CVD is a significant cause of morbidity and mortality in sickle cell disease [75]. CVD or stroke refers to a sudden onset focal or global neurologic deficit of vascular origin lasting more than 24 hours. It may be ischaemic or haemorrhagic. TIA or stroke occurs in 25%of patients with sickle cell disease. Overt stroke occurs in 10 to 15% of homozygous patients under the age of 10 years [85–87]. The prevalence of overt stroke among SCA children in PortHarcourt, Nigeria, is reported as 4.3% [88]. In Abuja, Nigeria, Oniyangi et al. reported a stroke prevalence of 5.2% among SCD children seen in a tertiary center [89]. The risk of CVD is higher in those with low baseline haemoglobin, low fetal haemoglobin, high white blood cell count, and high systolic blood pressure. CVD is rare in infants. Incidence of CVD is higher in Hb SS disease compared with SC disease, S/b+thalassemia, and S/b zero thalassemia [55, 78].

Infarctive CVD is commonest and occurs in patients aged less than 20 years and older than 30 years (peak incidence: 10–15 years) [90]. Infarction is often associated with stenosis or occlusion of affected vessels most commonly the distal internal carotid, proximal middle cerebral, and anterior cerebral arteries. The patient may present with antecedent history of TIA or seizures, which eventually progresses to an overt stroke, characterized by hemiparesis, speech, or visual impairment, or even coma if immediate therapy is not instituted. Suggested guideline on acute and long-term treatment of an ischaemic stroke in SCD is provided as follows.

Treatment Guidelines for Sickle Cell Ischaemic Stroke

(i) After initial evaluation of patient‘s airway, breathing, and circulation (ABC of resuscitation), further stabilization should be pursued through prevention and control of hypoxaemia, hypotension, hyperthermia, and glycaemic imbalance, which would worsen the cerebral insult.

(ii) Presence of seizures should be controlled with appropriate anticonvulsants. Prophylactic anti-seizure therapy is not necessary.

(iii) Urgent non-contrast CT/MRI is required to distinguish haemorrhage and infarction. This important distinction has to be made early, as this will impact subsequent therapeutic decisions.

(iv) In the early stage of brain ischaemia (<3 hours), crania lCT may be negative or show only subtle inconclusive signs. Magnetic resonance imaging (MRI) provides better details but should be deferred until treatment has been initiated.

(v) Early institution of exchange transfusion is crucial to improving treatment outcome. EBT should be targeted at reducing sickle Hb level below 30%. Simple transfusions may be offered in the interim while EBT is being planned. Simple transfusions at 10–15mL per kg red cells reduce sickle haemoglobin levels to about 60%.

(vi) Adequate hydration not more than 1.5 times the maintenance should be instituted with isotonic fluids preferably 0.9% normal saline.

(vii) In untransfused SCD patients, stroke recurrence rate is 67%, with 70% of recurrent strokes occurring in the first 3 years after the initial stroke [91]. As such, EBT should be followed up with hypertransfusion therapy to maintain sickle Hb level below 30% at a haemoglobin concentration of about 10 to 11 g/dL.

(viii) Chronic blood transfusion (CBT) has been shown to be beneficial in primary and secondary prevention of CVD [89, 92–94]. However, clear definitions on when and how CBT should be stopped is yet to be made. Often times, transfusions continue till late adolescence or early adulthood.

(ix) Hydroxyurea therapy reduces cerebral blood flow. Though less effective, hydroxyureamay be considered an alternative to chronic transfusion therapy, where transfusion is not feasible [86, 87].

(x) Thrombolysis with recombinant tissue plasminogen activator (rTPA) within the first 3 hours of ischaemic CVD in adult patients should be considered after careful patient evaluation [95]. TPA is not recommended in children. However, the current prospect of TPA use among Nigerian patients is remote due to challenges of its availability, cost, delayed diagnosis, and clinical experience with its use.

(xi) Antiplatelet agent, aspirin 325mg, is recommended if TPA is not used and should be avoided for the first 24 hours if TPA is used [96].

(xii) Adult SCD patients should be evaluated and treated for modifiable risk factors such as dyslipidaemia.

(xiii) Acute stroke should be treated in a dedicated stroke unit with input of both neurologist and haematologist.

Haemorrhagic stroke tends to occur between 20 and 29 years of age and is associated with low steady state haemoglobin levels and high steady state leucocyte counts [90].

Hemorrhage often results from rupture of vessels within the circle of Willis. Clinical presentation is similar to ischaemic CVD. However, patients with haemorrhagic CVDs are more likely to present with coma. In hemorrhagic CVD, patient may present with severe headache, vomiting, and other features suggesting raised intracranial pressure. Hemorrhagic CVD is rare but more fatal. Cerebral oedema is worse in hemorrhagic stroke. Prompt confirmation of diagnosis through imaging studies is required. Treatment of cerebral oedema with hypertonic solution such as mannitol is desirable. Antiplatelet and anticoagulants are contraindicated in hemorrhagic stroke. Though its exact role in hemorrhagic CVD is not clear, EBT is recommended especially for patients billed to undergo magnetic resonance angiography [95]. Surgical interventions by vascular surgeons may include ligation of accessible aneurysms and surgical vascular bypass procedure for moyamoya syndrome.

Nimodipine, a calcium channel blocker, improves outcome in adults with subarachnoid hemorrhage by counteracting delayed arterial vasospasm [95, 97].

Risk evaluation for an overt ischaemic stroke and the need for early preventive intervention are performed by assessment of cerebral blood flow velocity using transcranial Doppler (TCD) ultrasound. Cerebral blood flow in excess of 2 meters per second portends a high risk for CVD. Typically, TCD ultrasound assessment is commenced by age of 2 years. TCD between 1.7 and 2 should be reassessed in 3-4 months. If stable, assessment should be annual until 16 years of age [78]. Cerebral blood flow in excess of 2 meters/second is an indication for commencement of hyper transfusion therapy and this is shown to reduce stroke occurrence by about 90% [85]. MRI scan every 5 years may also be used in periodic evaluation of the brain for silent infarctions where resources are available [98].

Subclinical cerebral infarcts (SCI) in sickle cell disease patients occur in 27% and 37% of patients before their 6th and 14th birthdays, respectively [99]. Silent stroke is defined by an abnormal MRI in the absence of history and physical signs of an overt CVD. Risk factors for SCI include male gender, low steady state haemoglobin levels, higher baseline systolic blood pressure, and previous seizures [99].

Subclinical strokes are associated with neuropsychiatric dysfunction in apparently healthy SCD patients [98, 100] and are a risk factor for overt stroke [101]. In confirmed silent brain infarctions with neurocognitive delay and behavioural disturbances, chronic transfusion therapy is indicated.

Acute Chest Syndrome (ACS). ACS is a leading cause of mortality in sickle cell disease even among Nigerian patients, accounting for about 25% of all deaths [102–104]. Risk factors for acute chest syndrome includes older age, low fetal haemoglobin level, high haematocrit level, homozygous SS disease, chest VOC, smoking, general anaesthesia and surgery, asthma, and

possibly opioid use [105]. ACS is defined by new pulmonary infiltrates (on chest radiography) in at least one complete lung segment, fever, and at least one respiratory symptom (pleuritic chest pain, cough, dyspnoea, and tachypnoea) [105]. ACS may follow a painful crisis especially in adults. ACS may also complicate the immediate postoperative state. As such, there is need to maintain proper protocols for preventing or treating ACS during BPC and after surgery.

The underlying pathophysiology of ACS includes vasoocclusion of pulmonary vessels and microbial involvement. Implicated microbes include bacteria such as Pneumococcus, *Haemophilus influenza*, respiratory viruses, and atypical organisms such as *Mycoplasma*, Chlamydia, or *Legionella*. Respiratory viruses are more likely in children while bacterial causes are more frequent in adults [43]. Furthermore, hypoxia induced by ACS can trigger widespread sickling and vasoocclusion, with possibility of multi-organ failure and death. Fat laden pulmonary macrophages in the airways are observed in about half of the cases, suggesting possible contributions from bone marrow fat embolization [83]. Bone pain crisis involving the chest cage can also trigger ACS as a result of pain induced hypoventilation, which encourages sickling in the pulmonary bed and microbial growth. Similarly, oversedation with opioids may predispose to ACS.

During BPC or in the immediate postoperative period, care should be taken to prevent ACS. In such patients, prophylactic incentive spirometry is helpful [106]. In the absence of a spirometer, 10 deep breaths every 2 hours of the day is an alternative. Treatment of an established ACS also includes incentive spirometry/chest physiotherapy, parenteral broad spectrum antibiotics, effective pain relief, and supplemental oxygen therapy (2–4 liters per minute). Opioid is the mainstay of pain control in SCD. Its liberal use is encouraged in order to prevent pain undertreatment, prolonged treatment, and hypoventilation. However, care should be taken to avoid oversedation. Recommended antibiotic combinations include

quinolones, 2nd or 3rd generation cephalosporin alongside macrolides (for atypical bacteria). Antibiotic choice should be further directed by local susceptibility profile if available.

Bronchodilator therapy is also required as most patients may have a bronchoreactive component. EBT is indicated in worsening lung consolidation or persistent hypoxia, any neurological deficit (confusion, motor deficit, epilepsy), intractable pain or opioid intolerance, haemodynamic instability, nosocomial infections, acute worsening of anaemia or cardiovascular insufficiency, and acute enlargement of spleen or liver [78]. Mechanical ventilation is required in rapidly progressive cases. Inhaled nitric oxide and steroid may be helpful in life-threatening cases. Evidence suggests that recurrent episodes of ACS can be prevented by hydroxyurea [107]. Also chronic transfusion therapy is beneficial in secondary prevention of ACS and is indicated in patients with two or more episodes annually, who are unresponsive to hydroxyurea [108].

Priapism. Priapism is another acute complication of sickle cell disease. It is defined as persistent, purposeless, painful penile erection that is unassociated with sexual pleasure. Generally, reports of lifetime prevalence of priapism in sickle cell disease range from 2 to 35% [109]. Its prevalence is found to be as high as 44.9% among Nigerian male SCD patients [110]. Peak incidence occurs in 2nd and 3rd decade (median age: 18.5 years) [43, 110]. SCD priapismis a ―low-flow‖ type. The penile ischaemia results from outflow obstruction (poor venous drainage) caused by sickled cells. Usually, it affects the corpora cavernosa alone while the spongiosum is spared. A typical genital examination reveals a hard penis with soft glans; tricorporeal involvement is rare. The priapism may also be defined as stuttering, minor, or major (prolonged) depending on the duration of the attack and its frequency. Stuttering priapism typically last about 30minutes to 2 hours, tends to become recurrent (occurs several times a week), and may herald episodes of prolonged priapism.

Minor attacks occur infrequently or isolated. Major or severe attacks last longer than 3 to 4 hours and should be treated as a urological emergency. Severe (major) and recurrent priapism (penile ischaemia) is associated with irreversible organ damage, fibrosis, and impotence.

The goal of treatment is to preserve erectile function and prevent recurrences. As such, there is need for early presentation in the hospital if home remedies are unsuccessful within 2 hours of onset. At the onset, patients should be counseled to drink extra fluid, use home-based simple or compound analgesia, and attempt to void. Other self-help strategies such as warm baths and gentle exercises like jogging may be helpful. Oral dose of pseudoephedrine or terbutaline may be given. If the priapism persists more than 2 hours, hospital care is required. This includes intravenous hydration and opioid analgesia. If the priapism persists more than 3 hours, aspiration and irrigation of the corpora with dilute phenylephrine, epinephrine, or etilefrine is indicated. Frequently, aspiration of blood from the cavernosal bodies is performed with a 23-gauge sterile needle, followed by irrigation with a 1: 1,000,000 dilution of epinephrine in saline, after adequate counseling, conscious sedation, and local anaesthesia [111]. If detumescence is achieved lasting more than one hour, patient may be discharged home on oral analgesic, pseudoephedrine, and clinic follow-up. Penile aspiration and irrigation may be repeated up to 3 or 4 episodes if detumescence is not achieved early.

Simple early self-intracavernosal injection (SICI) of etilefrine and other adrenergic agonists such as metaraminol may achieve detumescence within one hour of onset, hence removing the need for hospital-based surgical aspiration and irrigation [112–114]. Sympathomimetics (adrenergic agonists) may be associated with untoward effects such as blood pressure changes and are yet to be licensed for SICI. EBT is indicated in recalcitrant cases. Surgical shunt procedures such as proximal shunt of quackel or distal shunt of winter may be tried, if conservative measures remain unsuccessful.

However, surgical penile shunts may also be unsuccessful and may induce impotence [115]. Often, priapism will resolve with one or a combination of medical interventions.

In preventing priapism, male sickle cell patients ought to be adequately informed and counseled about priapism from adolescence. Patients with frequent episodes (≥2 per month, ≥4 per year) should receive priapism prophylaxis with oral pseudoephedrine 30mg daily if they are less than 10 years of age and 60mg per day if they are older than 10 years. Etilefrine and diethylstilbestrol (DES) may be used prophylactically although evidence for its usefulness is limited [116, 117]. DES use has been limited by its feminizing effects, though a short course of 5mg dailymay be used to abort a stuttering episode [118]. Similarly use of injectable leuprolide, a GnRH antagonist, which works through endogenous suppression of androgen production, is associated with longstanding hypogonadism and rebound priapism after discontinuation [116]. Use of hydroxyurea may also be beneficial [116]. There is recent evidence that use of phosphodiesterase (PDE) inhibitor such as sildenafil is useful in preventing recurrent episodes of priapism [119–121]. Though not always successful, penile prosthesis may be remedial in those with established erectile difficulties persisting more than 12 months [122].

Ocular Disease. Central retinal artery occlusion by sickled red cell sludge is an ocular emergency. It manifests as sudden change in vision. It is treated like stroke. Treatment requires EBT, hyperoxygenation, and reduction of intraocular pressure with carbonic anhydrase inhibitors [43]. Prognosis is however poor.

Osteomyelitis. Osteomyelitis is one of the commonest skeletal complications of SCD [123, 124]. About 29% of Nigerian SCD patients experience this complication in their lifetime [125]. It often originates from bacteremia, as also observed in septic arthritis. Diagnosis may be quite difficult due to its similar presentation to acute bone infarction. Diagnosis requires a high

index of suspicion. Serial blood cultures, as well as culture of local bone aspirates, may be required [78].

The commonest cause of osteomyelitis in SCD population is *salmonella* spp, followed by *staphylococcus* species [43, 126, 127]. Treatment requires involvement of the orthopedic surgeon and clinical microbiologist. Broad spectrum antibiotic based on the common local isolates and their susceptibility profile should be commenced after culture samples have been taken.

1.6.Chronic Morbidities in Sickle Cell Disease

Delayed Growth and Development. Children with sickle cell disease have normal body weight at birth. However, by one year of life, there might be obvious weight lag when compared with normal infants. This weight deficit persists till adulthood and typically imparts a thin (asthenic) build [33]. Obesity may be seen in some cases [43]. Pubertal growth spurt may be delayed 1-2 years compared to their peers. Growth deficits in children with SCD may be due to multiple factors including severe anaemia, long-term effects of repeated vasoocclusion, endocrine failure, low dietary intake, and low socioeconomic status [128]. However, delay in skeletal maturation allows for bone growth such that final adult height is reached. Menarche may also be delayed for 1-2 years in females [62].

Chronic Pain Syndromes. There are two forms of chronic pain in SCD: chronic pain due to obvious tissue damage such as AVN or leg ulcers, and intractable chronic pain with no obvious cause. Suboptimal treatment of recurrent severe acute painful crisis may progress to an intractable chronic pain syndrome. There is need for prompt and adequate treatment of acute pain episodes. Opioids coupled with nonopioids and adjuvant remains the mainstay of analgesia in SCD [64].

Immunological and Infectious Complication. SCD patients have a subnormal immunity, which partly accounts for their increased susceptibility to infections [51]. Immunologic dysfunction in SCD is attributable to autosplenectomy with the resultant defective cellular and humoral immunity [129]. About 30% loss of splenic function occurs by first year of life and 90% by sixth year of life [130]. Normal splenic synthesis of immunoglobulins, properdin, and tuftsin is impaired, leading to increased susceptibility to infections. They are particularly susceptible to encapsulated organisms such as pneumococcus especially in children aged less than 5 years, hence the rationale behind pneumococcal vaccination and penicillin prophylaxis from four months of life till age five in western societies. Previous infections with *pneumococcus* confer lifelong prophylaxis. Studies reveal that, without preventive actions, invasive pneumococcal infection is 30 to 600 times more likely to occur in SCD children compared to normal persons [131]. *Haemophilus influenza* is the next most common organism and affects children older than 5 years. In Africa, *Salmonella, Klebsiella*, *Escherichia coli*, and *staphylococcus* seem to be more common than *pneumococcus* [131, 132]. As such, routine prophylaxis against *pneumococcus* is not an established practice in Nigeria [134]. However, there is recent compelling evidence from other parts of Africa that *pneumococcus* contributes significantly to infections in SCD [135–137]. Since infection has been documented as the commonest cause of death among SCD patients in Nigeria, the role of pneumococcus in SCD related infections and mortalities needs to be clarified through further research. There is a need for vaccination and chemoprophylaxis against common infections [102]. Current national immunization schedule in Nigeria routinely includes vaccinations against polio, tuberculosis, Diphtheria, tetanus, pertussis, hepatitis B, *Haemophilus influenza* infections, measles, and Yellow fever.

Before one year of life, the infant should have completed the vaccination schedule and is entitled to subsequent booster doses [138]. However, for persons affected with sickle cell

disease, additional compulsory vaccinations should be administered to cover for *Streptococcus pneumonia*, *Influenza virus*, and *Neisseria meningococcus*, *human papillomavirus (HPV)*. Children less than two years of age should have four doses of the 7 valent pneumococcal vaccine between 2 and 15 months of life. The 23 valent pneumococcal vaccine should be administered at age of 2 years and older and should be repeated every 3 to 5 years till 10 years of age and every 5 years for those older than 10 years. Influenza vaccines should be administered during cold seasons beginning at 6 months of life. HIB vaccine should be commenced at 2 months of life. Meningococcal vaccination is recommended for patient at 5 years and older and is repeated every two years. HPV vaccine is administered to females under 26 years.

Also contributing to increased risk of infection in sickle cell disease is repeated tissue infarctions, which are potential foci for pathogens. Similarly, iron overload in patients that have had several transfusions favors growth of iron dependent bacteria such as *Yersinia enterocolitica*. Furthermore, micronutrient deficiency especially zinc deficiency is associated with lymphopenia and decreased immunity [139]. About 60–70% of SCD patients are zinc deficient. In Nigeria, a case control study showed the serum zinc level to be significantly lower among SCD children compared with healthy controls [140]. Other studies have also shown significant deficiencies of other micronutrients such as magnesium and selenium [141, 142].

Sickle Cell Chronic Lung Disease. Sickle cell chronic lung disease (SCCLD) is an age related morbidity. It affects at least a third of adult SCD patients [78]. Patterns of the lung involvement among Nigerian patients include restrictive lung disease, obstructive lung disease, chronic hypoxaemia, and pulmonary hypertension (PHT) [78, 143–145]. A Study in Nigeria reported a prevalence of 18.9% for SCCLD among adult SCD patients [146]. Chronic complications occur more frequently in those with history of acute chest syndrome.

In about 20% of patients, echocardiography shows elevated pulmonary artery systolic blood pressure >35mmHg. Incidence of PHT is higher in patients with high haemolytic rates and high LDH. PHT is associated with 10-fold increase in the relative risk of death and it confers poor prognosis [147]. Primary PHT is not the only cause of elevated TRV (tricuspid regurgitation jet velocity) in SCD patients. Patients with elevated TRV have increased risk of mortality in the next 3 years [78]. Treatment includes hydroxyurea therapy, chronic transfusion, vasodilator use, anticoagulation, and oxygen therapy.

Hepatobiliary Complications. Chronic liver damage in sickle cell disease is caused by intrahepatic trapping of sickle cells, transfusion transmitted hepatotropic infections, and transfusion siderosis [78, 148]. Evidence suggests that post-transfusion hepatitis and other transfusion transmissible infections are still a significant problem in Nigeria [149, 150].

In Ibadan, Nigeria, Fashola and Otegbayo observed a post-transfusion viral hepatitis prevalence rate of 12.5% in 2002 [149]. In rare instances, vasoocclusion in the liver with cholestasis may precipitate acute liver failure. Pigment gallstones are found in about two-thirds of sickle cell patients, especially those with sickle cell anaemia [148]. Symptomatic gallstones require cholecystectomy. Cholecystitis is treated with antibiotics.

Treatment of asymptomatic cholelithiasis may require watchful waiting. Gall stones associated with common bile duct stones require endoscopic retrograde cholangiopancreatography (ERCP). The risk of hepatic damage is reduced by ensuring viral safety of all transfused blood components and prompt institution of iron chelation therapy if iron overload is present. In hepatic failure, liver transplantation is a veritable option.

Other Abdominal Complications. Incidence of peptic ulcer disease (PUD) is higher in SCD patients. PUD occurs in about 35% of SCD patients with epigastric pain [98]. In Ile-Ife, Nigeria, Akinola et al. observed PUD among 28% and 50% of Hb SS and Hb SC disease

patients presenting with abdominal pains, respectively. Interestingly, duodenal ulcers are not associated with high acid outputs; rather, ulcers are secondary to decreased mucosal resistance, possibly due to bowel ischaemia and NSAID abuse.

Renal Complications. The hypoxic, acidotic, and hypertonic state of the renal medulla favors vasoocclusion and destruction of the vasa recta. By the first year of life, SCD infants may develop hyposthenuria manifesting as nocturia or enuresis [62]. Local studies have shown that the prevalence of nocturnal enuresis is higher among children with homozygous sickle cell disease [151]. This further makes them susceptible to dehydration, especially in hot climate.

In addition, distal type IV tubulopathy in SCD promotes acidosis, further predisposing to vasoocclusive events. Papillary necrosis (usually of the left kidney) presents with haematuria. Other possible causes of haematuria include infections, stones, and tumor. Recommended guidelines for treatment and prevention of sickle cell nephropathy (SCN) are presented as follows.

Recommended Guidelines for Management of Sickle Cell Nephropathy

(i) SCN is an age-related morbidity. Among Nigerian patients, its prevalence and severity increases with advancing age, longer survival, and homozygous SS disease [152, 153].

(ii) Relevant clinical history and examination findings such as facial puffiness, loin pain, painless haematuria, leg and abdominal swelling, frothy urine, worsening anaemia, and hypertension, which may suggest renal disease, should be elicited at regular intervals during visits.

(iii) At least once annually during maintenance visits, SCD patients should be assessed for their renal status. Recommended laboratory assays include urinalysis (on every visit), serum electrolytes, urea and creatinine (semiannually), creatinine clearance/estimated

glomerular filtration rate (eGFR), and tests for microalbuminuria (albumin creatinine ratio, ACR; urinary protein to creatinine ratio, uPCR). A normal creatinine level does not exclude renal disease in SCD due to supranormal kidneys precipitated by hyper-filtration and increased secretion of creatinine and uric acid. Emphasis and therapeutic decisions should be placed on significant adverse changes in the renal markers rather than single absolute values.

(iv) Consultations and co-management with experienced nephrologist is recommended in the following setting: patients with uPCR >50mg/mmol (442mg/g), persistent microscopic haematuria, declining renal function (>10% fall in eGFR per annum), or eGFR <60 mL/min/1.73m2. Further evaluations including renal biopsy are necessitated in settings of sudden onset heavy proteinuria with or without nephrotic syndrome [154, 155].

(v) Treatment of haematuria includes bed rest, hydration, and blood transfusion if indicated in events of a significant blood loss. Most times, haematuria is caused by papillary necrosis. However, the possibility of a renal medullary cell carcinoma must be excluded in these patients.

(vi) Progression of SCN to ESRD is often heralded by worsening proteinuria, anaemia, and hypertension. This may be delayed with adequate control of hypertension and proteinuria. Introduction of angiotensin converting enzyme inhibitors (ACEIs) or angiotensin receptor blockers (ARBs) reduces proteinuria [136]. For blood pressure control, diuretics are better avoided.

(vii) Similarly, early commencement of hydroxyurea helps to delay progression to ESRD except hydroxyurea is contraindicated for other reasons.

(viii) NSAIDS for pain control are better avoided in patients with established SCN, in order to prevent worse organ damage. NSAIDS cause significant decline in renal blood flowand glomerular filtration.

(ix) Urinary tract infection in these patients should be treated aggressively. Patients with ESRD should be on regular EBT especially if renal transplant is being planned. End stage renal disease is managed with repeated dialysis, erythropoietin therapy, and/or renal transplant.

Ocular Disease. Incidence of ocular disease is higher in Hb SC disease, Hb SB+thal compared to Hb SS [78, 156]. Repeated vasoocclusion in the vascular beds of the eye especially the retina causes progressive ophthalmopathy, which manifests as comma-shaped conjunctival vessels, iris atrophy, retinal pigmentary changes, and retinal hemorrhages. Neovascularization leads to sea-fanning, so called proliferative retinopathy. Eventually, vitreous haemorrhage and retinal detachment may occur. For prevention, annual eye examination is recommended for all SCD patients from the 2nd decade of life [78]. Treatment options for proliferative retinopathy include laser photocoagulation and vitrectomy.

Sickle Cell Leg Ulcer. Leg ulcers are frequent in adults SCD patients especially males with AS phenotype and patients with low steady state haemoglobin levels [157, 158]. In a report from Benin City, Nigeria, Bazuaye et al. observed a prevalence rate of 9.6 and 22.4% for current ulcers and previous ulcers, respectively [158]. Ulcers commonly arise near the medial or later almalleolus and may be single or multiple. The aetiology is often multifactorial and they include vasoocclusion of skin microvasculature, made worse by trauma, infection, warm climate, and iron overload[43]. Commonly isolated microbes include *pseudomonas aeruginosa*, *staphylococcus aureus*, and *streptococcus* species. Chronic SCD ulcers are painful and resistant to healing. Treatment of these ulcers requires multidisciplinary approach involving the haematologist, plastic surgeon, specialist nurses, and orthopaedic surgeon [43].

Generally, treatment includes pain relief (including local pain control before wound dressing), elevation of the leg, debridement (to remove necrotic tissue), elastic dressing/support bandage, and zinc sulphate therapy (600mg/day). Some patients may benefit fromchronic blood transfusion and skin grafting.

Musculoskeletal Complications. Known musculoskeletal complications in SCD include medullary hyperplasia, dystrophic intramedullary calcification, H-vertebra, osteolysis, osteopenia, septic arthritis, dactylitis, ulcers, pathologic fracture, and osteomyelitis. H-vertebra or Cod-fish vertebra is due to infarction of the vertebral body, giving a fish-mouth appearance on radiography. In a study by Balogun et al., musculoskeletal complications occurred in 31.4% of adult Nigerian SCD patients [159]. Avascular osteonecrosis of the femoral and humeral head is particularly associated with reduced quality of life. AVN develops in about 50% of patients who survive to above 35 years of age and about 60% of patients who survive to 60 years of age [160]. Another recent study revealed that AVN occurred in about 13 per 1000 Nigerian SCD patients [161]. Exact mechanism for development of AVN is yet to be clearly described. Even patients with high fetal haemoglobin levels may not be totally protected from developing AVN. However, high steady state platelet count has been correlated with AVN in Nigerian SCD patients [161]. Other clinical and laboratory correlations of AVN include high haematocrit, coexistence of alpha thalassemia, and frequent

VOC [162–164].

In older patients, humeral head necrosis is more common than femoral head necrosis, although femoral head necrosis is associated with more devastating pain due to weight bearing [165]. Treatment of musculoskeletal complications of SCD requires comanagement with an orthopedic surgeon with special interest in SCD. Persisting pain in a joint or at least a stage 3 arthropathy is indication for referral to an orthopedic specialist [165]. X-ray features of AVN may not be obvious until repair processes have changed the density of the bone. MRI is

the investigation of choice in SCD patients with persisting hip or shoulder pain. Every patient with confirmed AVN should be staged with MRI. Initial conservative treatment should include counseling/patient education, analgesia, partial weight bearing on crutches, and physiotherapy. Option of joint replacement/arthroplasty is available for patients with severe joint destruction.

Cardiovascular System Changes. Sickle cell disease is associated with cardiac abnormalities including dilated cardiomyopathy, ventricular hypertrophy, cardiac iron overload, dysrhythmias, pulmonary hypertension, myocardial infarction, and sudden death [166, 167]. Chronic anaemia in SCD potentiates ventricular hypertrophy and dilatation which may progress to left ventricular diastolic dysfunction and exercise intolerance [166]. Pulmonary arterial hypertension is defined by end systolic pressure in the right ventricle greater than 25mmHg (normal is less than 15mmHg). In a cohort of Nigerian SCD patients, 2 (3.6%) out of 56 met the criteria for pulmonary hypertension [144]. Tricuspid regurgitation jet velocity of >2.5m/sec is associated with a high risk of pulmonary hypertension and is an independent risk factor for death [147].

Transfusion Related Morbidities. Blood transfusion is a key therapeutic modality in SCD. In a cohort of Nigerian SCD children, the prevalence of blood transfusion is as high as 57% [168]. Benefits of transfusion in sickle cell disease include correction of the baseline anaemia, dilution of sickle haemoglobin levels, and suppression of endogenous sickle red cell production, as well as reduction in chronic haemolysis and circulating sickle cell levels [169–171]. Transfusion modalities in SCD include simple transfusions, exchange blood transfusion, or chronic blood transfusion (hypertransfusion).

Simple transfusion refers to top up correction of anaemia. Indications for chronic blood transfusion include prevention of first stroke, prevention of repeat stroke, TCD USS >2m/sec,

delayed growth and development in children, frequent ACS, severe disease, severe SCD lacking HLA match, sickle chronic lung disease, pregnant women with bad obstetric history and frequent bone pains, and sickle cell leg ulcers [169, 170]. Indications for exchange blood transfusion include moderate to severe ACS, refractory painful VOC, stroke, central retinal artery thrombosis, and acute refractory priapism[55].The choice of blood component for transfusion in SCD should be a sickle negative, recently donated (less than 7 days old), leucodepleted, and phenotypically matched for at least Rh and Kell antigens, racial and minority matched red cell concentrate. Cytomegalovirus (CMV) negative component should be used for transfusion in all CMV negative children, as they may be candidates for bone marrow transplantation.

Target haemoglobin level should not exceed 10- 11 g/dL in SCD as there are concerns for hyperviscosity and vasoocclusion [45, 172].

Transfusion of blood and blood components is not without risks. In particular, delayed haemolytic transfusion reaction and alloimmunisation are among the immunologic complications of blood transfusion associated with sickle cell disease. Due to their tendency for repeated transfusion from chronic prophylactic transfusion or otherwise, the risk of iron overload in body tissues with irreversible organ damages ensues, hence the need for close monitoring and prompt iron chelation when indicated. Reports on alloimmunization rate among Nigerian SCD patients are lacking and may be related to the lack of routine alloantibody screening and extended red cell phenotyping in most blood banks in Nigeria.

Among non-immunologic complications of transfusion therapy in SCD in Nigeria, transmission of viruses and iron overload is of note [108, 150]. Recent evidence suggests that transmission of viruses is still a major challenge to transfusion safety in Nigeria [150]. This calls for a better national transfusion service. SCD patients, particularly those on hyper-

transfusion therapy, are at particular risk for iron overload, with resultant damage to vital organs [108]. Iron status should be monitored in SCD patients, particularly those who have received a cumulative transfusion dose of more than 20 to 30 units. Chelation therapy should be instituted promptly if serum ferritin levels exceed 1000 ug/L [173].

Psychosocial Issues/Psychiatric Complications. Psychosocial complications of SCD include poor self-image, negative thoughts and feelings about the condition, stigmatization, depression, cognitive impairments, fears, anxieties, hatred for parents and others, dropping out of school, and tendency for substance abuse [41, 174]. These complications are associated with the chronic nature, recurrent pain, reduced health related quality of life, and unpredictable course of the disease. Some degree of psychologic trauma is also rendered to parents and health caregivers. A recent report by Anie *et al*. revealed that about half of Nigerian SCD patients had depressive feelings [175]. Adequate psychological support should be provided for patients by physicians, other health care staff, parents, and support groups. Those requiring more definite intervention should be co-managed with clinical psychologists and psychiatrists.

A study in Jamaica revealed that 29% of SS patients had a psychiatric disorder, compared to 25% in the control population [176]. Association of psychiatric morbidity included leaving school early, difficulties in social adjustment, impaired cognition, and previous psychiatric difficulties [176].

Asthenic body builds and abnormal facies may be associated with poor self-image. Such patients should be identified early and treated. Other complications such as under-education and underemployment may require the services of medical social worker and occupational therapy unit.

1.6.1. Special Care Situations

Sickle Cell Disease and Pregnancy. Typically, pregnancy in female SCD patients is attended by anaemia which may be worsened by pregnancy related plasma volume expansion and folate deficiency. VOC is more common in 3rd trimester [177]. Increased incidence of preeclampsia, maternal mortality, and perinatal complications such as abortions, stillbirths, low birth weight, and neonatal deaths are associated with SCD pregnancy [177–179]. As such, pregnant SCD patients require special care by specialists including experienced obstetrician, haematologist, midwives, and anaesthesiologist. Preferably, oral contraception should be recommended for sexually active SCD females and pregnancy should be planned. Folate supplementation should be ensured. A local study has shown that preconceptual care and early antenatal booking produce better outcome and less obstetric complications in SCD pregnancies [180]. Hyper-transfusion is indicated in cases of bad obstetric history and severe sickle cell disease in pregnancy. SCD and pregnancy are procoagulant states. Coupled with obstetric surgeries, the risk of VTE is significantly increased and appropriate anticoagulation may also be necessary in their care-plan [64].

Perioperative Care in Sickle Cell Disease. Perioperative complications of surgery in SCD patients include hypoxia, dehydration, bone pain crisis, significant anaemia, and acute chest syndrome [181]. Anaesthesia may be associated with hypoxia and dehydration [182]. Good anaesthetic expertise and experience is indicated when undertaking surgical procedure in SCD patients [182]. Early surgical complications such as pain and haemorrhage should be well controlled. Optimal analgesia and tact surgical skills are indicated.

Other strategies to improve perioperative outcomes in SCD include conservative preoperative blood transfusion therapy, epidural analgesia, and adequate postoperative pain control with

opiate and nonopiate analgesia [183, 184]. Aggressive or exchange transfusion therapy has not been associated with better surgical outcomes [181, 183].

Sickle Cell Disease and Radiology. Infusion of radiologic contrast media may precipitate VOC. Hypertonic nature of contrast media triggers marked intracellular dehydration and marked increment in red cell MCHC, thus precipitating sickling. This complication may be averted by preprocedure red cell exchange to achieve target sickle haemoglobin level of 50%. Traditional iodinated contrast media (due to its high osmolality) are relatively contraindicated in SCD. Isotonic contrasts are safer to use in SCD [185].

1.6.2. Laboratory Diagnosis of Sickle Cell Disease

The science behind laboratory diagnosis of sickle cell disorder entails phenotypic testing for the presence the sickle haemoglobin and genetic analysis. Physicochemical properties of the sickle haemoglobin such as decreased solubility and sickling under deoxy conditions, its pattern of mobility in an electric field, and rate of elution from solution unto adsorbents are applied in its laboratory detection. Phenotypic tests may be used as screening tests or diagnostic tests. Screening tests chosen for the purpose of mass screening should be highly sensitive and cheap to run. Examples of screening tests include sickling test, solubility test, and alkaline haemoglobin electrophoresis. On the other hand, high specific, diagnostic tests include isoelectric focusing, citrate agar electrophoresis, and high performance liquid chromatography [186, 187].

Quantification of haemoglobin variants and globin chain studies are used in evaluation of compound heterozygous disease states such as sickle thalassemia syndrome [186, 188]. Hb A2 levels in excess of 3.5% are suggestive of haemoglobin S-beta thalassemia [187]. Other ancillary laboratory investigations useful in detection and monitoring of the disease include FBC, reticulocyte count, and peripheral blood film. Reticulocyte count usually range from 5

to 15% in sickle cell disease. On peripheral blood film examination, findings may include irreversible sickled red cells, polychromasia, occasional nucleated red cells, and schistocytes, as well as Howell-Jolly bodies [41, 130]. Target cells are seen in sickle haemoglobinopathies. In sickle cell thalassemia syndromes, target cells are seen alongside microcytes and moderate-severe hypochromia. Red cell indices may suggest macrocytosis due to increased reticulocytosis or compliance with hydroxyurea therapy. However, oval macrocytosis with hyper-segmented neutrophils may suggest folic acid deficiency.

Biochemical changes include high LDH, low haptoglobin, high total and indirect bilirubin, and high AST [64]. Genetic studies such as PCR are used for prenatal and pre-implantation diagnosis [189].

1.6.3. Prognosis and Life Expectancy

Severe SCD is associated with poor outcomes, if no intervention is rendered. Known modulators of clinical severity include fetal haemoglobin levels, beta globin haplotype, amd coinheritance of alpha-thalassemia, as well as geographical and other unknown genetic factors [34, 35]. In a study by Emmanuel Chide et al., a higher leucocyte count was associated with more SCD complications in a Nigerian SCD population [190]. Another recent Nigerian study in a cohort of 115 children with SCD showed the presence of dactylitis at first presentation and higher total WBC, neutrophil count, platelet count, and serum bilirubin levels to be significantly higher among those with severe disease, while a higher fetal haemoglobin level was associated with a milder disease [191].

Other notable poor prognostic factors include low haemoglobin F production, Hb less than 7 g/dL, Hb greater than 7 g/dL, high VOC rate, pulmonary HTN, and nocturnal hypoxaemia (more strokes) [78].

From a large cooperative study in USA in 1994, the median survival for SCA was reported as 42 and 48 years in men and women, respectively. For haemoglobin SC disease, it was reported as 60 years and 68 years for men and women, respectively [103]. In USA, 95% of children with SCD survive till adulthood [192]. In Jamaica, survival estimates for persons with SCA were reported as 53 years and 58.5 years for men and women, respectively [193].

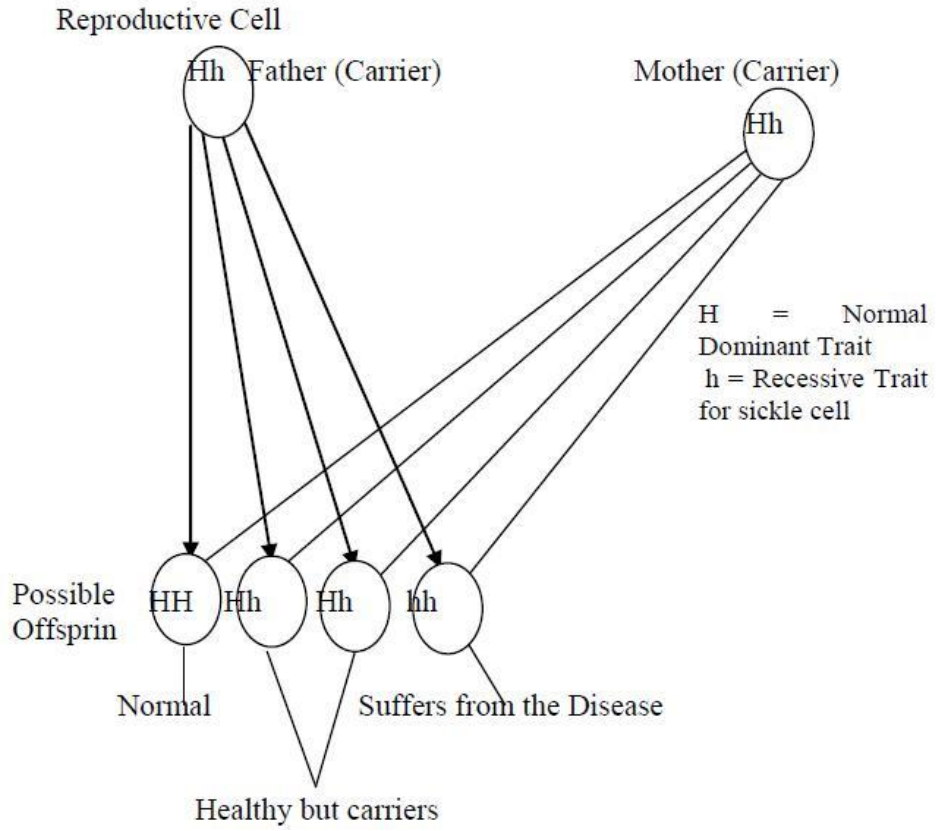
Life expectancy in SCD is substantially reduced especially in those with severe disease. In a 10-year retrospective study reported in 2009 from Ilorin, Nigeria, by Chijioke and Kolo, the mean age of sickle cell anaemia patients was found to be 23 years compared to 40 years in the control population, suggesting reduced life expectancy [194]. Findings from that study also revealed that age correlated negatively with survival [194]. As recently reported by Ogun et al., the leading causes of mortality in Nigerian SCD patients include infections, acute chest syndrome, anaemia, acute sequestration crisis, and stroke. According to the study, the mean age at death was 21.3 years. Though some patients now attain fifth decade, most mortality occurs in their second and third decades of life [102].

1.7.The Role of Social Welfare Counselling in the Eradication of Sickle-Cell Disease in Nigeria

Midence and Elander[195] asserted that Sickle cell is a disorder of the blood. Part of the blood are the red cells because the blood contains plasma, which is the fluid with, tiny particles that are invisible to the naked eyes, but are actually living cells. Red blood cells, the white blood cells and the platelets are millions in the blood. Inside this red blood cell are substances called haemoglobin which carry oxygen from lungs to the different parts of the body. Because the blood circulates, this oxygen is eventually carried to the body and is released to make them function. In the affected individuals, the red blood cells are sickle, that is, they take on distorted shapes and become rigid. When sickle red corpuscles become wedged in the

capillaries, blocking local blood flow, the sickle cell haemoglobin molecules occur, forming pseudo crystalline structures known as tactoids; this process is known as sickling. The sickle cell disorder is hereditary, not infectious. The disease has been traced to a single recessive gene. Because the gene is recessive, it must have been inherited from both parents for a child to develop the disease [196]. Any child with one sickle cell gene and one normal gene in his or her genetic makeup is completely healthy, but he or she is a carrier; that is she/he carries the sickle cell trait, and capable of passing the trait on to his/her offspring. If a sickle cell carrier marries someone who also has the sickle cell gene, some of their children may develop the disease and some of them may not. The chances of children inheriting sickle cell disease where both parents are carriers can be illustrated by the diagram below.

Hereditary possibilities of transmitting sickle cell diseases



Counselling and prevention of causes and infections are simple measures not readily accessible to most patients. As a result, the majority of children with the most severe form of

the disease die before the age of five, usually from an infection or severe anaemia. The survivors remain vulnerable to exacerbations of the disease and the complications mentioned above.

SCD has major social and economic implications for the affected child as well as the family. Recurrent sickle-cell crises interfere with the patient_s life, especially with regard to education, work and psychosocial development. Presently, there is no cure for SCD. However, cost-effective treatment exists for the pain and other aspects of the disease. The most important components of this treatment are early intervention with analgesics, antibiotics, rest, good nutrition, folic acid supplementation and high fluid intake. At times, invasive procedures such as blood transfusions and surgery may be needed.

SCD is present mostly in blacks. It is also found, with much less frequency, in eastern Mediterranean and Middle East populations. Individuals of Central African Republic descent are at an increased risk of overt renal failure. The sickle gene is present in approximately 8% of black Americans. The expected prevalence of sickle cell anaemia in the United States is 1 in 625 persons at birth. The actual prevalence is less because of early mortality. More than 2 million people in the United States, nearly all of them of African American ancestry, carry the sickle gene. According to National Institutes of Health and Centres for Diseases Control and Prevention; the following statistics are available:

- ✓ Sickle cell anaemia is the most common inherited blood disorder in the United States.
- ✓ More than 70,000 people in the United States have sickle cell disease.
- ✓ Sickle cell disease occurs in 1 in every 500 African Americans.
- ✓ About 8% of African Americans are carriers of sickle cell disease.
- ✓ Two million people have sickle cell trait.
- ✓ Approximately 1 in 12 African Americans has sickle cell trait.

In the United States, SCD accounts for less than 1% of all new cases of end-stage renal disease (ESRD) [197]. The following factors are known to portend a greater likelihood of progression to overt renal failure; hypertension, nephritic-range protein-uria, severe anaemia, and a Central African Republic heritage [198]. In patients with SCD, 5 – 10% developed renal failure [199]. In 2006, the World Health Organization (WHO) pronounced Nigeria as the country with the highest number of suffers of sickle cell anaemia in the world. The global health watch along put the annual number of sickle cell anaemia sufferers in Africa at about 200,000, noting that Nigeria account for 150,000 sickle cell anaemia children every year [200]. In Nigeria, there is no accurate account since the majority born to rural dwellers does not survive childhood[201]. He ascertained in the research that about 9000 births would have sickle cell disorder by the end of that year. However between 1992 and till date, the number has greatly increased because of multiplication effects and due to Nigerians flair for children. Unfortunately, the issue of Malaria has complicated and worsens the issue. Most people with normal blood genotype – AA, die prematurely due to severe illness caused by malaria. Infants and children are vulnerable because they would have had fever infections and could not build up immunity against the parasite. Hence, the available AS genotype inter-marry which makes majority become carrier of SCD; the carriers would then increase in number making the control more difficult.

However, recent medical findings [202] show that about 30 percent of Nigerians are carriers of the mutant gene, with the prevalence rate at 20 per 1,000 births. ―With the carrier frequency ranging between 20 percent and 30 percent of Nigerians population, it means that more than 30 million Nigerians are carriers‖ the report stated. The disease, according to medical sciences is inherited from both parents and is usually caused by some abnormalities in a type of haemoglobin called haemoglobin-s. Symptoms of the disease vary, but a research conducted by Adeyokunnu and Hendricks in Sheyin[200] both of the University College

Hospital, Ibadan, shows the sufferers have pain episodes, usually referred to as crisis which can last between hours and days. They asserted in their research that some carriers suffer abdominal pain, breathlessness, delayed growth and puberty, fatigue, fever, ulcers, among others.

Senator Gyang Dantog, Chairman, Senate Committee on Health, notes that sickle cell anaemia has been proved to be more fatal than HIV/AIDS. He describes the effects of the disease as very devastating and declared that Nigeria could not afford to remain its ―World Capital. Dr Artemos Francis, a Kaduna-based medical practitioner, believes that the first step toward checking the menace is to prevent the occurrence. He asserted that sickle cell can only occur when two people, who carry the sickle cell trait, have a child together. Hence, it is necessary to intensify counselling intending couples to go for tests to determine their status. The federal government of Nigeria showed concern in the struggle to abate the burden sickle cell patients do experience. In his speech while commissioning the Multi-million Naira Ultra-Modern National Sickle Cell Centre, opposite Lagos University Teaching Hospital (LUTH), Idiaraba Lagos, the then President, Olusegeun Obasanjo expressed his worry over the sickle cell anaemia incidence in Nigeria and asked the Committee to advise the Federal Government on what to do to reduce the scourge of the disease in the country. In his address the Chairman of the foundation, Professor Olu Akinyanju (2007) disclose that, Nigeria by her large population harbours the highest number of sickle cell patients in black Africa, with 150,000 new patients yearly and 40 million people with the traits of the disease.

1.7.1. Complications and Problems Associated With Sickle Cell Disease

According to University of Maryland, Medical Centre[203], there is still no cure for sickle cell disease other than experiment transplantation procedures but treatments for complications of sickle cell have prolonged the lives of many patients who are now living into adulthood. The

hallmark of sickle cell diseases is the sickle cell crisis which is an episode of pain. It is the most common reason for hospitalization in sickle cell disease. In general, the risk for a sickle cell crisis is increased by any activity that boosts the body‘s requirement for oxygen such as illness, physical stress or being at high altitudes. In more than half of episodes, however, the trigger is unknown. Episodes typically begin at night and last 3 – 14 days, accelerating to a peak over several days and then declining. Pain most commonly occurs in the lower back, leg, hip, abdomen or chest, usually in two or many locations. Pains in the bones are common because blood obstruction can directly damage bone and because bone marrow is where red blood cells are manufactured. Acute Chest Syndrome (ACS) occurs when the lung tissues are deprived of oxygen during a crisis. It can be very painful, dangerous and even life threatening. It is a leading cause of illness among patients with sickle cell disease and is the most common condition at the time of death. The pain often lasts for several days. In about half of patients, severe pain develops about 2 – 3 days before there are any signs of lung or chest abnormalities. Acute Chest Syndrome is often accompanied by infections in the lungs, which can be caused by viruses, bacteria or fungi. Pneumonia is often present.

Infections are common and an important cause of severe complications. Before early screening for sickle cell disease and the use of preventive antibiotics in children, 35% of infants with sickle cell died from infections. Fortunately with screening tests for sickle cell now required for newborns, and with the use of preventive antibiotics and immunizations in babies who are born with the disease, the mortality rate has dropped significantly. Such infections pose a serious threat to infants and very young children with sickle cell disease. They can progress to fatal pneumonia with devastating speed in infants, and death can occur only a few hours after onset of fever. Infections are also common in older children and adults with sickle cell disease, particularly respiratory infections such as pneumonia, kidney infections and osteomyelitis, a serious infection in the bone.

About 30% of patients with sickle cell permanent partial or complete erectile dysfunction can occur. Enlargement of the liver occurs in over half of sickle cell patients and acute liver damage occurs in up to 10% of hospitalized patients. Because sickle cell patients often need transfusions, they are at higher risk for viral hepatitis, an infection of the liver. This risk, however, has decreased since screening procedures for donated blood have been implemented. Gallbladder disease is common among sickle cell patients. About 30% of children with sickle cell disease have gallstones, and by age 30, 70% of patients have them. In most cases gallstones do not cause symptoms for years. When symptoms develop patients may feel overly full after meals, have pain in the upper right quadrant of the abdomen, or have nausea and vomiting. Acute attacks can be confused with a sickle cell crisis in the liver. Ultrasound is usually used to confirm a diagnosis of gallstones. If the patient does not have symptoms, no treatment is usually necessary. In some children with SCD, excessive production of blood cells in the bone marrow causes bones to grow abnormally, resulting in long legs and arms or misshapen skulls. Sickling that blocks oxygen to the bone can also cause bone loss and pain. Sickling that affects the hands and feet of children causes a painful condition called hand foot syndrome. A condition called avascular necrosis of the hip occurs in about half of adult sickle cell patients when oxygen deprivation causes tissue death in the bone. Eventually adult patients may need surgery to remove diseased and dead bone tissue. Patients with severe causes may need joint replacement.

Leg sores and ulcers may occur in the sufferers of SCD. They usually affect patients older than 10 years. SCD can also damage blood vessels in the eye and cause scarring and detachment of the retina, which can lead to blindness. Women with SCD who become pregnant are at higher risk for complications such as miscarriage and premature birth, and their babies may have low birth weight. SCD symptoms often worse during pregnancy and pain crises become more frequent. However with careful prenatal care and monitoring,

serious problems can be avoided. Older children and adult patients with SCD are subject to other medical problems, disease have pulmonary hypertension. Stroke is the second most common killer of patients with sickle cell disease who are older than 3 years old. Between 8 – 10% of patients suffer strokes, typically at about age 7. Strokes are usually caused by blockages of vessels carrying oxygen to the brain. Patients with sickle cell disease are also at high risk for strokes accused by aneurysm, a weakened blood vessel wall that can rupture and haemorrhage. Multiple aneurysms are common in sickle patients but they are often located where they cannot be treated surgically. Anaemia is a significant characteristic in sickle cell disease commonly referred to as sickle cell anaemia. Because of the short lifespan of the sickle red blood cells, the body is unable to replace red blood cells as quickly as they are destroyed. This causes a particular form of anaemia called haemolytic anaemia. Most patients with sickle cell disease have haemoglobin levels of about 8gldL, much lower than healthy people. Chronic anaemia reduces oxygen levels and increases the demand on the heart to pump more oxygen bearing blood through the body. Eventually, this can cause the heart to become dangerously enlarged, with an increased risk for heart attack and heart failure.

The kidneys are particularly susceptible to damage from the sickling process. Persistent injury can cause a number of kidney disorders, including infection. Problems with urination are very common, particularly uncontrolled urination during sleep. Patients may have blood in the urine, although this is usually mild and painless and resolves without damaging consequences. Kidney failure is a major danger in older patients and accounts for 10 – 15% of deaths in sickle cell patients. About 40% males, including children with SCD suffer from priapism. Priapism causes prolonged and painful erections that can last from several hours to days.

There was a caption, _pain, pain and pain‘ in the widely read News Paper, _Sunday Punch‘ of 24th September 2006. This was an expression of some Nigerians, parents of sickle cell disease patients who met and lamented on the agony they go through and the discrimination that goes with it. In a rare display of emotions, many of them narrated the pain they have had to suffer for harbouring a dysfunctional genotype. Some of the women who spoke with *Sunday Punch* however claimed that they were not ignorant of the disease, but were victims of wrong and faulty laboratory diagnosis.

1.7.2. Symptoms of Sickle Cell Disease

Sickle cell disease is usually diagnosed during childhood. However, some milder form of the disease can be missed if certain blood tests are not complete. One should be concerned about having sickle cell disease if one has:

i. Unexplained pain in abdomen, chest, back, joints and muscles fatigue.
ii. Anaemia that does not respond to iron supplements.
iii. Family history of sickle cell disease or sickle cell trait.
iv. Medical Laboratory Test.

If one has been diagnosed with sickle cell disease, one can decrease the frequency of pain crisis by following some simple guidance.

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

Another treatment given to sickle cell anaemia patients is Bone Marrow Transplanting (BMT). It is being reported with increasing frequency but remain controversial in spite of encouraging results. The type of BMT done for sickle cell anaemia patients is Alogenic Bone Marrow Transplantation. The concern raised by opponents to this treatment for sickle cell anaemia are the acute and long term complications, morbidity, and costs compared with traditional therapy of disease Mirabel tea is also used to boost Haemoglobin levels among people who have anaemic conditions such as SCD. Mirabel tea has tremendous anti-sickling properties. Regular consumption of Mirabel tea has been associated with the prevention of the crisis and pain in people living with SCD. There is also a control measure, which is the termination of early pregnancy. The foetus is tested at the embryonic stage, and if found to be a carrier, the pregnancy is terminated at the early stage before three months. However, the woman is subjected to carrying many pregnancies in which only few that are tested to be non-carriers will be allowed to live. As a matter of fact, this is also expensive and it can endanger the life of the woman in question. The best option to embark on is total eradication.

End Notes

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Chapter Two

2. Social Work: Conceptualization, Goals, Ethics and Values

Social work is a practice-based profession that promotes social change, development, cohesion and the empowerment of people and communities. Social work practice involves the understanding of human development, behavior and the social, economic and cultural institutions and interactions. Social work is a problem-solving profession that is committed to improve the quality of human life by imparting various intervention techniques and strategies which are scientifically proved and artistically applied[1].

Social workers are professionals who aim to enhance overall well-being and help meet basic and complex needs of communities and people. Social workers work with many different populations and types of people, particularly focusing on those who are vulnerable, oppressed and living in poverty. Social workers are educated and trained to address social injustices and barriers to their client's overall wellbeing. Some of these include poverty, unemployment, discrimination and lack of housing. They also support clients and communities who are living with disabilities, substance abuse problems or experience domestic conflicts[2].

Social Work has been recognised across the globe since inception as a Human Service Profession and like many of its western counterparts - the profession of social work has gained a global recognition and has paved the way for social change across the globe through its approach of charity and now evolving to an empowerment model. From charity to professional social work and social welfare, this profession has evolved though the trajectory from - welfare; to development; to social justice, to rights based approach and as an empowering profession[3].

Indeed the dominant ideology of Social Work as a profession has evolved from a medico-socio thematic perspective to dynamic rights based empowerment approach catering to a multiple issues and problems.

Social work is a problem-solving profession that is committed to improve the quality of human life by imparting various intervention techniques and strategies which are scientifically proved and artistically applied. Social work is a profession primarily concerned with the remedy to psycho-social problems and deficiencies which exists in the relationship between the individual and his social environment[4]. This phenomenon always existed in the society in one form or the other, but achieved its scientific basis in the last decades of 19th century.

People in distress, destitution and deprivation have been helped in the past by individuals joint families, caste communities, religious institutions motivated by religious philosophy that assumed _charity‘ as a reward or medium for salvation, humanism, philanthropy, humanitarian feeling, democratic ideology, equality of all citizens; regard for human personality; respect for other‘s rights including the indigents, the handicapped, the unemployed, emotionally challenged and above all, those in need[5].

The charitable work taken up by individuals and some voluntary organizations came to be termed as social work in the due course of time. The most frequently seen misconception about social work is that people mistake charity, alms giving and voluntary activities as social work, but this in reality is not social work, as it does not provide any permanent solution to the person‘s problems and does not empower a person to face his problems by himself[6]. Social work aims at making a person self-dependent and self-reliant. It attempts to explore the casual factors behind the problem and tries to solve them scientifically.

The social life of the modern world is characterized by numerous problems. The consumeristic and profit-oriented approaches of the present society bring about drastic consequences in social life. Problems like isolation, poverty, unemployment, migration, family disintegration, sexual violence, atrocities against women, suicide, substance abuse, communalism, terrorism, child abuse etc. are on the increase. Solution to these problems on a scientific basis is essential to ensure peaceful social life. Many people give alms to solve problems like poverty and unemployment[7].

We have been practicing these types of charity for centuries to help the poor and vulnerable, still the problems exist. These activities provide only temporary solutions to their problems. The complexity and magnitude of such problems in modern times demand systematic approaches. Professional social work makes people independent rather than dependent. When it was observed that the problems require scientific approaches for the proper and effective management, a new system of scientific intervention came into existence, that ism social work[8].

Basically, social work is a helping activity which adopts scientific and systematic approaches to address the issues of individuals, groups and communities and it empowers the society. It is the only profession which studies social problems, its origin, interrelation etc. and provides systematic solution to such problems. Hence, the present-day social life demands a scientific and professional approach to solve social problems[9]. Social work, as a profession and scientific discipline, is of recent origin. Common men are not aware of the significance of social work. A Social Worker is a trained person who receives remuneration for his services like any other professions. Social work is a helping activity which aims at enabling individuals, families, groups and communities to become self-reliant[10].

Social work is based on certain principles and philosophy that help to practice it with scientific knowledge and skills. It requires education and training for any person to practice it[11]. Therefore, it is visualized that social work is scientific process for helping to the needy suffering from material deficiencies, physical disabilities, mental disorders, emotional disturbances and so on by using knowledge in human relationship and methods and skills in dealing with them.

In a nut shell, social work intended to assist individuals, families, social groups and communities in sorting out their personal and social problems and permanently solving those problems through a systematic process[12].

1. Social work is a professional and academic discipline that seeks to improve the quality of life and well-being of an individual, group or community.
2. Intervention at individual, group and community level is undertaken in social work.
3. Relationship is the key tool of social work.
4. It promotes study, diagnosis, research and resource mobilization.
5. Social work simply means _help people to help themselves‘.
6. We can introduce social work through the Chinese proverb "Give a man a fish, and you feed him for a day; show him how to catch fish, and you feed him for a lifetime‖. This proverb means that equipping/preparing someone for a job is of greater benefit than a one-off hand out[13].
7. Social work in its various forms addresses the multiple, complex transactions between people and their environments.
8. Its mission is to enable all people to develop their full potential, enrich their lives, and prevent dysfunction.
9. Professional social work is focused on problem solving and development.

10. Social workers are agents of change in society and in the lives of individuals, families and communities they serve. Social work is an interrelated system of theory and practice[14].

Social work as ―an art of bringing various resources bear on individual, group and community needs by the application of a scientific method of helping the people to help themselves‖ [15]. In his definition Stroup gave a different look to social work saying it an art. He said that it is an art of bringing the people into the position where they themselves will be able to solve their problems. Social work is a welfare activity based on humanitarian philosophy, scientific knowledge and technical skills for helping the individual, group or community to live a rich and full life[16].

Social work is a practice-based profession and an academic discipline that promotes social change and development, social cohesion, and the empowerment and liberation of people. Principles of social justice, human rights, collective responsibility and respect for diversities are central to social work. Underpinned by theories of social work, social sciences, humanities and indigenous knowledge's, social work engages people and structures to address life challenges and enhance wellbeing[17]. The above definition may be amplified at national and/or regional levels.‖ Social work is ―a profession devoted to helping people function the best they can in their environment.

2.1.The Goals of Social Work

The ultimate goal of social work is to enhance the well-being and level of functioning for all people and to create positive social change by improving social conditions and creating more humane practices and policies for vulnerable populations.

1. The goal of social work is to reduce suffering by solving people's problems. People have psychosocial problems with regard to their physical and mental health. Apart from this, adjustment problems-in children and adults can be dealt with separately.
2. Social work enhances social functioning of individuals, groups and families by providing recreational services to the public, and by a judicious use of leisure time, can prevent delinquency and crime in the society[18].
3. Social work helps the individual in bringing about a change in the environment in favour of his growth and development.

   Social work provides democratic ideas and encourage the development of good interpersonal relations, resulting in proper adjustments with the family and neighbourhood.

1. It works for social justice through legal aid
2. It also promotes social justice through the development of social policy.
3. Social work improves the operation of social service delivery network as well.

2.2.The Functions of Social Work

Function of social work refers to the natural activity of social work or the statements of how social work operates. Social work operates to assist individuals in adjusting to the institutional frame work of the society and attempts to modify the institutional frame work itself in appropriate areas[19]. He classified the functions of social work into the following four major categories:

1. Curative Function

The services provided under curative functions are–medical and health services relating to psychiatry, child guidance, child welfare services, services for the handicapped or disable in

the form of protection and rehabilitation[20]. These kinds of services aim to cure the physical, social, material, psychological sickness of individuals in the society.

2. Correctional Function

The correctional function of social work has three broad areas, such as:

a) Individual reform service which includes prison reform, probation, parole and other related services.

b) Services for improving social relationship which includes family welfare services, school social work, industrial social work etc.

c) Services for social reform that includes employment services, prevention of commercial sex work, beggary prohibition services and removal of untouchability[21].

3. Preventive Function

It includes life insurance services, public assistance, social legislation, adult education and prevention of diseases etc. This type of function basically deals with the services relating to the prevention of problems like insecurity, unlawfulness, ignorance, sickness etc. It is directed towards the elimination of those factors in the social environment or those deficiencies in the development of personality that prevents the individual from achieving a minimum desirable standard of socio-economic life[22].

4. Developmental

Developmental function includes the tasks of socio-economic development activities such as: education, recreational services, urban and rural development programmes and programmes of integration etc. which are primarily concerned with the development of individuals,

families, groups and communities. The basic functions of social work can be divided in three broad interdependent and interrelated categories, viz;

3. Restoration of social functioning
4. Provision of resources and
5. Prevention of social dysfunction.

The restoration of impaired social functioning is the oldest and most known function of social work profession. This function is subdivided into curative and rehabilitative aspects. The curative aspects are to eliminate environmental factors that have caused break down of social functioning of individuals, groups or communities and the role of the rehabilitative aspects is to recognize and rebuild interaction patterns in the society[23].

As mentioned earlier, social work tries to intervene at the point where the individual interacts with his environment. The environmental factors hindering the functioning of a person may be social, economic, political or cultural. This function emphasizes at problem solving through modification in the psycho-social environment of individual and groups and through bringing about changes in the attitude of recipients of the services[24]. Therefore, if a person becomes dysfunctional due to any of the above-mentioned environmental factors, the first task at hand will be to restore the person‘s normal functioning. The second task will be to assess the damage caused by the dysfunction and strategies are devised to rehabilitate the person socially and bring him back to the normal mainstream.

Provision of Resources is further subdivided into developmental and educational. The developmental aspects are designed to extend effectiveness of existing social, human and material resources or to bring about full utilization personal capacity for more social interaction. The educational functions are designed to make the public aware about specific conditions and needs for new and changing resources and approaches[25].

The prevention of social dysfunction involves early detection, control and elimination of conditions and situations that could obstruct effective social functioning. The main two divisions are preventions of problems in the area of human interaction (individuals and groups) and secondly, prevention of social ills. Although it is a very important function of social work, it is neglected in most situations. Social work has generally concentrated on the curative and rehabilitative function and has worked on the problem-solving model[26].

However, keeping in the mind the rapidly changing social scenario, it has become imperative to adopt the preventive approach to social work. The profession should ensure that problems are neither created nor eliminated at the very beginning. For this function the social workers play a crucial role of conscientization, capacity building and organizing people so that they themselves can prevent social dysfunctions[27].

In the present-day context, social work has to emphasize on the aspects of change. It is increasingly being realized that the main cause of dysfunction lies not with the people but with the systems within which they operate. Either the people do not get and environment or resources required for proper social functioning or they do not have access to the resources to fulfill their needs. Therefore, there is a need to challenge and change the system so that people get a favourable environment to function. In a nutshell, it can be said that the function of social work should shift from a status quo service oriented approach to change oriented development approach[28].

2.3.The Ethics and Values of Social Work in Nigeria

Ethic within the context of social work is defined as the moral principles concerning right and wrong that guides one‘s behavior whereas, it was viewed as a ―moral philosophy… that involves systematizing, defending, and recommending concepts of right and wrong conduct.‖ In the other hand, _values‘ is defined as ―a person‘s standards‖ in life. Both ethics and values

are sometimes used interchangeably. However, it is noteworthy to establish the fact that, while ethics represent the moral philosophy guiding human behavior, values set the minimum standards for moral behavior. Therefore, it is clear that human behavioural attitudes are usually guided by both values and ethics. So, to this end, there can be personal ethics and institutional ethics, as well as individual values and institutional values[29].

It was explained the ethics of the Social Work Profession in Nigeria as follows; Social workers must be transparent in dealing with their clients, they should be ready to keep as confidential, the secret entrusted to them, they must be people of good integrity. Social workers should promote the right and interest of their clients, respect the diversity of cultural beliefs, promote the independency of service users; not to exploit the concerned party, avoiding neglect of their clients and not using abusive words to their clients; self- composure, knowledge ability, proper documentation, record-keeping as well as the establishment of a healthy relationships with the clients[30].

He added that, in his own case as a social worker, he always made sure that he never took chances of any of his clients. He believed that, the service users are depressed and need comfort and empowerment, not exploitation. On the other hand, a scholar states that, his professional values are the readiness to impact great change in peoples‘ life. He emphasized that respecting peoples‘ cultural dispositions and religious beliefs is a value that brings about peaceful co-existence[31].

Social Welfare Administration and Social Work

The field of social work is the academic study of coordinating social services for vulnerable populations. Social work, social group work, and community organization are three of its most significant scientific and research subfields. Face-to-face engagement with clients is the focus of social casework, which as a branch of social work science is concerned with the

evaluation, diagnosis, documentation, analysis, and resolution of clients' issues via the application of relevant theories, principles, and practices[32]. The focus of social casework is on addressing each client's unique set of problems. It has been around since the beginning of social work and is still in use today.

As the second broad category of social work, social group work is an increasingly important subdiscipline. Social group work focuses on clients by helping them see the value of group dynamics. Many professionals in the field of social work hold the view that a group of people working together to tackle a common issue has more success than any one person working alone. While both social group work and social casework rely heavily on social workers' codes of ethics, the latter use methods that are unique to themselves[33].

The third subfield of social work is community organization, which takes a more systematic approach to addressing people's needs. When planning for the betterment of a community, it's important to think about the impact any given action could have on a wide cross-section of the populace. Community organization, like casework and social group work, is a method used in social work to reach out to individuals who are in need of government aid or support[33].

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Chapter Three

Sickle Cell Disease Control and Current Challenges in Nigeria

Control of SCD begins with public education and definite strategies to prevent further transmission of the trait. Carrier detection and genetic counseling have been proven to be successful in curbing the spread of other haemoglobinopathies like thalassaemia [1]. Carrier detection should be offered at designated centres after proper genetic counselling through antenatal and newborn screening, couple/premarital screening, and other forms of population screening. Genetic counseling by trained personnel helps individuals at risk to take informed decisions about their reproductive life choices. The option of prenatal diagnosis and selective abortion in Nigeria is controversial and relatively unavailable. Local studies show that a significant proportion of Nigerians are averse to selective abortion, even if legally permitted [2–4]. Early detection and diagnosis of sickle cell disease is crucial to reducing mortality and mortality associated with sickle cell disease, as affected persons are offered early supportive and preventive treatments. Despite the huge burden of SCD, currently, there is lack of national or regional SCD newborn screening programme in Nigeria, as at the time of this publication.

Specialized centers dedicated to care of SCD patients with requisite multidisciplinary teams and other facilities are grossly absent. Ideally, SCD infants diagnosed prenatally or through newborn screening should be routed to comprehensive SCD centers for optimal treatment [5]. Conversely, Nigerian SCD is still associated with delayed diagnosis [6].

Continuous training of healthcare professionals involved in care of SCD patients is also desirable. Further efforts should be directed at education of the patients and their parents or caregivers. The health caregivers should also constantly undergo professional refresher and update courses in order to optimize their knowledge and skills in care of SCD. Recent

surveys still suggest a dearth of public health knowledge on sickle cell disease in Nigeria [7]. Despite Nigeria being the most populous black nation on earth with the highest burden of sickle cell disease, till now, there are no coordinated nationwide efforts aimed at controlling the disease. Current evidence suggests that the care available for patients with SCD in Nigeria is still suboptimal [8]. Secondary control measures such as chronic transfusion therapy and use of hydroxyurea are faced with peculiar challenges in developing nations such as Nigeria [9]. Such challenges include unavailability of blood and blood components, the need for patients and relatives to regularly source for blood and blood donors, cost of iron chelation, risk of transfusion transmissible infections, and overall cost of chronic blood transfusion [10, 9]. A recent study estimated the mean annual cost of hypertransfusion in Ibadan among paediatric SCD patient to be 3,276 US Dollars (SD = 1,168) [10]. Also, treatment of iron overload with metal chelators, which is a potentially inevitable complication of chronic transfusion, increases cost.

Furthermore, HSCT, which is the only potentially curative disease modifying intervention in sickle cell disease, is currently available in Nigeria and has been reported [11]. However, its practice is bewildered by ample challenges including poor government commitment, weak political will, poor infrastructure, unaffordability by the average eligible Nigerian SCD patient, lack of local bone marrow registries, absence of specialized molecular diagnostic laboratories, and epileptic electric power supply

[12, 13].

3.1. Psychological Impact of Sickle Cell Disease

SCD, and especially the pain episodes, can have widespread impact on both the psychological functioning of the individual diagnosed with the illness and their families. Children with SCD are at risk for maladjustment in almost every area of daily functioning [14, 15]. Specifically, SCD has been associated with several indicators of psychological

maladjustment including emotional and behavioral problems, poor self-concept and interpersonal functioning, limited athletic abilities (due in part to illness restrictions), and poor academic performance[16, 17, 18].

With respect to the family, caregivers of children with SCD are burdened with missed work, increased family stress, and increased disease care demands, which is due in part to the unpredictability of pain crises care in SCD[19]. Caregivers of children with SCD are tasked with the responsibility for managing their child's care, which includes encouraging their child to engage in preventative behaviors, managing pain episodes, teaching coping skills, and providing adequate nutrition/ hydration. Moreover, the primary caregivers often report a lack of support by family and friends during the child's pain crises, which contribute to feelings of hopelessness, helplessness, and frustration[20].

Health Related Quality of Life

Research has demonstrated that there is a unique interplay between the patient's psychosocial adjustment and the pathophysiology of SCD[21]. Given the increase in medical advancements and subsequent decreases in disease morbidity and mortality, more attention has focused on quality of life (QOL), which is an individual's assessment of his or her satisfaction with various aspects of his or her life (e.g., physical, emotional, school, social). Health related quality of life (HRQOL) refers more specifically to the impact of the child's illness on their subjective well-being[22]. Measuring QOL has become increasingly important for its function in evaluating interventions, assessing prognostic factors, comparing therapies, and allocating resources[23].

Although there are few studies, findings suggest that QOL outcomes in children with SCD are generally poor[24, 25]. For example, Kater et al. [25] found that a pediatric population with SCD had lower daily functioning and general physical limitations than parents of healthy

children. In 2002, Palermo et al. found that children with SCD were experiencing more psychosocial maladjustment compared to the healthy controls. Moreover, children with SCD had significantly more limited general health and physical functioning, more limitations in their academic functioning and social activities attributed to their physical health, and more behavior and emotional problems when compared to a healthy control group. Moreover, in an adult SCD population, research has shown that the frequency of sickle cell pain episodes over a 12 month period was associated with impairment in QOL [26].

With approximately 10-20% of children reporting frequent and severe SCD-related pain [27], it is likely that recurrent pain negatively impacts QOL. Research has shown that, SCD pain predicts several facets of QOL including school absences [28], lower academic performance [29], decreased participation in social activities [30], and sleep disruption [31]. Further, the frequency of these pain episodes is associated with decreases in QOL [28, 26]

Panepinto et al. [24] examined the role of SES and disease severity in pediatric SCD. The criteria used to define disease severity included SCD-related complications (i.e., stroke, acute chest syndrome, hospitalizations for VOC's, and recurrent priapisms) and interventions (i.e., hydroxyurea or bone marrow transplantation). Panepinto et al. [24] were interested in clarifying the compounding nature of impoverishment in the African American community on reports of QOL. Participants included 104 children with SCD and 74 healthy controls aged 2-18 years. Results supported prior research documenting the negative effects of disease severity on QOL in children with SCD [15, 26, 32]. Overall, children with the poorest QOL were those with a higher disease severity (e.g., frequent pain), older age, and more economically disadvantaged families. Given the debilitating effects of SCD pain, it is likely that the frequency with which children experience VOC's will greatly impact their QOL.

Chronological Age

Research has shown that developmental status has a significant influence on psychological adjustment in pediatric populations [33, 34]. Previous research has found the relation between caregiver distress and child maladjustment in pediatric chronic illness to be interactional in nature[35, 36]. Thus, it is important to consider the role of developmental status on caregiver and child adjustment. Adjustment to chronic illness is influenced by individual, family, and illness factors [37]. Typically, a gradual shift in disease-related responsibility from parent to child occurs over the course of development [38] such that greater autonomy and independence are anticipated. This change in roles and responsibilities can actually lead to increases in both stress and conflict in both the adolescents and caregivers [39]. Research in other pediatric chronic illness populations has found that decreases in parental involvement is associated with poor disease management in adolescents with cystic fibrosis [40], diabetes [41], and HIV [42]. Therefore, older children with a chronic illness may actually require greater parental assistance and less autonomy [43] than healthy comparison adolescents. Caregiving demands which are inconsistent with normative development have been associated with greater distress especially among parents of older children [44, 45]. Over time, the prolonged stress of raising a chronically ill child may impact caregiver adjustment. Aside from the additive effect of attending to multiple stressors, it is likely that caregiver‘s expectations of their child‘s ability to meet age-related milestones will influence their subjective experiences. Of note, it is important to consider that whereas age may be indicative of developmental expectations, it is not synonymous with biological indices of child development. Thus, in light of the relation between parental age-related expectations and their own adjustment in other chronic illness populations, it will be important to explore the impact of developmental status in pediatric SCD.

Caregiver Adjustment

Caregivers of children with a chronic illness often report increased parental distress and negative affect [46]. Thus it is not surprising that caregivers of children with SCD have alarmingly high rates of maladjustment. For example, previous research has consistently found that approximately 24% of parents of young children with SCD report clinically significant levels of psychological distress; similar to that of adolescents with SCD [47]. In tandem, [48] found that 35% of the primary caregivers in the sample met criteria for clinically significant levels of poor adjustment and 65% were at risk. The rates of psychosocial maladjustment in parents of children with SCD are similar to the high rates found in individuals diagnosed with a medical condition (20%-25%; [49]).

Bachanas et al. [50] found that parental psychosocial maladjustment was a significant predictor of their child's psychosocial maladjustment. Research has shown that the general outcome for caregivers of a child with a chronic illness is similar to that of the child [51]. Moskowitz et al. [52]found that in addition to time providing crisis care, parents of children with SCD are spending a greater proportion of time in disease-related technical care such as medication administration and diagnostic procedures than parents of a child with HIV. Furthermore, possibly due to the unpredictability of pain crises, caregivers of children with SCD are also reporting greater care burden [52]. Fifty percent of the caregivers of children with SCD in this study were at risk for clinical depression compared to 34% of parents of children with HIV.

Although there is limited research examining the relation between SCD pain and parental psychosocial adjustment, there is ample research documenting this relation in pediatric chronic pain[53]. Given that SCD is a chronic illness characterized by both acute and chronic pain, it is likely that the chronic pain literature can be applied to the population under study. Though rare, as of late more attention has been directed towards the families of children

experiencing pain [54]. Specifically, parents of children with chronic pain are not only directly impacted by the stress of parenting a chronically ill child; they are influential in both the child's adjustment to and experience of the pain [53]. Consistent with the research on parents of children with SCD, research with parents of children with chronic pain has shown that these parents often report high levels of anxiety and depression

[55, 56, 57].

Jordan, Eccleston, and Osborn [57] utilized qualitative methodology to examine the caregiver's experience of parenting an adolescent with chronic pain. Jordan et al. [57] conducted focus groups with 17 parents of adolescents (age 11-18 years) with chronic pain recruited from two tertiary care clinics in the United Kingdom. The researchers utilized Interpretive Phenomenological Analysis (IPA), a qualitative analytical method, for exploring the data. Two themes emerged which were labeled the caregiver's ―struggle for control and coherence‖ and a ―very different life‖. Moreover, these caregivers highlighted the disruption in developmental milestones which contributed to their feelings of uncertainty. Though there are disease-related differences, these themes parallel the difficulties regarding unpredictability which is associated with SCD VOC's.

Palermo and Chambers [54] examined the role of parent and family factors associated with pediatric chronic pain and disability. Palermo and Chambers[54] argue that there has been a lack of an integrative contextual framework for adequately conceptualizing the effects of parenting-specific behaviors and broader family factors on child pain. Their model highlights the reciprocal relationship between child pain and parental factors (e.g., psychosocial adjustment). Palermo and Chambers[54] suggest that researchers design future studies based on integrative theoretical models which will contribute to our understanding of the multifaceted nature of pediatric pain.

3.2. The National Burden of Sickle Cell Disorder and the Way Forward

Molineaux and co-workers[58], after some research in northern Nigeria, correctly summarized the situation when they wrote in 1979, ―*There is no other known inherited disorder present at such high frequency in a large population and of comparable severity as sickle cell anaemia in Africa. With rising standards of living and control of malaria, sickle cell anaemia will become an immense medical, social and economic problem throughout the continent*.‖ In sheer numbers, Nigeria has the largest burden of sickle cell disorder (SCD) in the whole world. Carriers of the sickle cell gene (Hb AS) have, over the past centuries, flourished and multiplied in tropical sub-Saharan Africa because their carrier status protected them from succumbing to the deadly falciparum malaria prevalent in the Region. In other words, they enjoyed a survival advantage over their peers who had not inherited the gene (Hb AA) and those who had inherited it from both parents and therefore had sickle cell anaemia (Hb SS).

Although countries around Nigeria also have an S gene carrier frequency of about 1 in 4 of their populations, Nigeria's large population has ensured that over 40 million Nigerians are healthy carriers of the S gene. This number of carriers far exceeds the total population of every other affected African country and indeed, of several of them put together. Consequently, about 150,000 Nigerian children are born each year with sickle cell anaemia (HbSS), the prevailing type of sickle cell disorder (SCD) in this Region. Survival of these children beyond childhood is largely dependent on their access to appropriate care and as most of them are born into poor underprivileged families, very few of them survive childhood[59]. On the other hand, the survival of those with access to good care, at all ages, is steadily improving although many challenges to their quality of lives and life spans still remain.

The Level of Awareness.

For an old disorder with a birth rate of 1 in 50 babies, the level of awareness is relatively low. This is not because of a lower birth rate previously, but because the affected children rarely survived childhood and were therefore less likely than now, to be encountered in secondary schools, in universities and in the workplace. Their peers and relatives now have the opportunity to observe their periodic pain crises and, sadly, occasionally, their premature deaths. This familiarity has heightened awareness. The other factor boosting awareness is the development of Sickle Cell Clubs, with attendant publicity, over the past two to three decades.

Response to the National Burden of SCD Despite its 2% birth incidence[59], the estimated population of SCD affected persons in Nigeria is only about one million, owing to a high rate of premature deaths. Unfortunately, the increasing awareness has not been matched by the development of a well-resourced national policy. This has curtailed the dissemination of accurate and unbiased public information and education about the disorder and has fuelled the growth of myths, misinformation, inappropriate treatment, frustration and stigmatization. The frustration has kindled the desire in many Nigerians to *do something about sickle cell disorder*. What needs to be done often appears deceptively easy and is usually not fully thought out and remains a subject of confusion and controversy even among health care workers.

In this regard, one frequently hears talk of eradication of the disorder while the wider context of management and control is invariably overlooked.

Selective Mating and the Eradication of SCD

Eradication of SCD by selective mating appears so logical and simple that many individuals, religious bodies and charitable organizations have tried to implement it themselves by screening young people and instructing those shown to be carriers to avoid choosing spouses who also have the sickle cell gene. Some churches even refuse to marry them.

This strategy is based on the false assumption that SCD can be easily eradicated by mass population screening of haemoglobin genotypes as a basis for coercive selection of spouses, The Military Administrator of Oyo State, had, in 1995, proposed a punitive edict aimed at prohibiting marriages between carriers of the sickle cell gene, but the conference of Solicitor– Generals in Nigeria thwarted its introduction by declaring that it was unconstitutional and offended the human rights charter to which Nigeria is a signatory[60].

The reality however, is that enforced selective mating of couples has never been shown anywhere in the world to have reduced the incidence of any inherited disorder. Attempts to introduce it by the Church in Cyprus, for the control of thalassaemia (also an inherited haemoglobin disorder), there, led to increased anxiety and stigmatisation of affected persons and of healthy carriers of the gene[61, 62]. This, in turn, led to widespread denial and falsification of haemoglobin genotype results among carriers who wanted to marry each other.

What the Church in Cyprus now does is to ensure that all couples have been counselled on thalassaemia before marriage.

Another popular but false assumption, is that, all marriages between carriers of the S gene are contracted in ignorance. As health workers in the specialty, we in fact see many marriages contracted between carriers who are both well-educated and informed about their Hb status,

and between them and persons affected with sickle cell anaemia. I know at least two Nigerian doctors who have married their SCD affected patients. One of the doctors was himself a carrier of the S gene. Although I knew the other doctor quite well I did not bother to ask whether he was also a carrier, as that would have seemed intrusive and perhaps, even judgmental. A few marriages contracted between affected individuals (e.g. SS and SS; SS and SC) have also been reported in Nigeria and elsewhere.

A research study of the effect on choice of marriage partner of informing young people and their families of their carrier status was carried out in the Arta area of Greece, where 20% of the population carries either thalassaemia or sickle cell gene. All young people of marriageable age were screened and counselled, and counselling contact was maintained for a two-year period. When the pattern of marriages was assessed at the end of the period, screening had had no measurable effect on choice of partner [63].

These examples simply go to show that there is more to marriages than Hb genotypes. We must ask ourselves whether we have got the moral right to choose spouses for others or simply to allow them to make informed choices.

Given our present state of knowledge, eradication of SCD can only be feasible by the unthinkable genocide of all healthy carriers of the gene or their enforced banishment from the country!

Stigmatization

Stigmatization and discrimination are products of ignorance and insensitivity. They are not only felt and encountered by affected persons and their families, but also, to a lesser degree, by carriers of the S gene. Some examples may blow your minds.

A secondary boarding school student stopped taking his daily medication because his schoolmates derided him for doing so. He was too ashamed to inform them of his condition and need to take daily medication.

A University of Lagos lecturer with two affected children himself was trying to organise a Sickle Cell Club on the campus for the benefit of affected students.

He publicised his mission asking for affected students to come forward. Many did not do so, for fear of identification and stigmatization. Nobody wants to be different or be regarded as an object of pity, or feel by implication of widely expressed ―solutions‖ that his birth was an unwanted mistake which should not have happened if only both parents had known better and avoided marrying each other. *_What a pity that these people were born. Let us inform everyone to avoid marrying so called ―incompatible spouses‖ who can cause the births of others like the‖*. Some of these statements and sentiments expressed by their peers are, of course, well-meaning, but, on reflection, highly insensitive and borne out of ignorance of some realities. So, the affected student feels like an unwanted alien and would like to hide in denial. You can imagine the resulting loss of confidence and self-esteem.

A young Nigerian woman was devastated when the overbearing father of her fiancée prevented their marriage on discovering that, she, like his son, was a carrier. She told us that, since then, she had changed her Hb genotype to AA.

In early May 2009, the Punch Newspaper reported that a 10 year old girl was found starved and sleeping on the streets in Yenagoa, Bayelsa State. Some kind-hearted people rescued her and learnt that when her parents separated she was taken by her mother. When her mother went to live with her new boyfriend, he rejected keeping her as soon as he found out that she had sickle cell anaemia. She was ejected, even though her father had assured her mother that he would continue to bear the cost of her maintenance and education.

While still trying to fathom that cruelty to an innocent child, a friend of mine in Lagos told me that he had offered to foster, accommodate and pay all the living and educational expenses of the 7 year old son of a recently deceased friend.

Another friend who was inexplicably winking at him while he was offering to do so later told him why he was trying to attract his attention. It was to warn him against fostering the boy whom he knew had sickle cell anaemia.

The cruelty factor notwithstanding, the last two cases would suggest that many people are not aware that people with SCD can live long useful lives, and, in this

country, are found in all the usual civic professions as well as in politics, where, I know at least two, who have risen to the positions of State Governor and State Commissioner respectively.

Marketing of Worthless Remedies

Charlatans and other adventurers have, not unexpectedly, flooded the market with drugs that are worthless to people with SCD. These drugs have been condemned by the Nigerian Society for Haematology and Blood Transfusion (see their Communiqué on page 44 of The Punch of August 28, 2008). Many people buy these drugs because they bear NAFDAC registration numbers on their packages. When in 2008 we met Professor Dora Akunyili, the former Director-General, she insisted that NAFDAC registration was meant only to assure the public of the safety of the drug for human consumption and is not an approval of its efficacy in the treatment of the condition or disease for which it is advertised. This is hardly understood by members of the public and the Sickle Cell Foundation Nigeria is still in discussion with NAFDAC over this.

Multiplicity of Sickle Cell NGOs

Diverse sickle cell NGOs, some merely duplicating rather than consolidating efforts, and others, worryingly with dubious motives and unclear capacity, have started to emerge. Many are driven by an emotional over simplified vision of eradication without training in genetic counselling or in any aspect of its management. This tendency, if unchecked will disseminate mixed messages that can confuse members of the public and thus become counter-productive.

3.2.1 The Way Forward

Coordination and Harmonization of Institutions and Programmes

The Sickle Cell Foundation Nigeria (SCFN) was established to address the problem of SCD in Nigeria in a systematic, scientific and sustainable manner. It has achieved its first goal of developing, in accordance with WHO recommendation [64], a comprehensive National Sickle Cell Centre in Lagos. The intention is for each State to have a collaborating Sickle Cell Centre that will supervise and coordinate the programmes and activities of all Sickle Cell Clubs within that State.

Training and Capacity Building

The National Sickle Cell Centre is engaged in training various cadres of health care personnel in order to improve the standard of preventive and curative care available to persons with SCD.

3.3. Prevention of SCD: Counselling as a Panacea

Genetic Counselling

Genetic counselling is not marriage counselling. Sickle cell disorder should be sensitively managed within the community. Otherwise, people living with SCD or who are healthy

carriers of the gene may feel stigmatised and be tempted to conceal, deny or falsify their haemoglobin (Hb) genotype status. This state of affairs can be counter-productive to efforts aimed at managing the disorder in the community. The importance of genetic counselling in preventing these undesirable outcomes is well established.

In counselling, the client is a person with SCD, his close family member or a healthy carrier of the sickle cell gene or the gene of some other haemoglobin variant such as Hb AC. The role of the counsellor is to ensure that the clients are given accurate unbiased information necessary to assist them to reach their own decisions on reproductive behaviour or any other course of action related to the disorder. Their decision must be respected and supported by the counsellor and confidentiality must be maintained. Failure to do so usually alienates the client to the detriment of the relationship between counselor and client.

Effective counseling is informative, confidential, non-directive and supportive. It requires skills that are best acquired through appropriate training and experience. Doctors who see many patients with SCD should endeavour to acquire the necessary skills and, if they have many patients, employ the services of one or more trained nurse-counsellors.

3.3.1. The Principles of Genetic Counselling

Informative

Genetic counseling should provide accurate and unbiased information about the disorder. To do this effectively, it is necessary to find out the prior knowledge and perceptions of the client concerning SCD. Any myths, wrong or superstitious beliefs should be uncovered, corrected and then replaced with accurate information. Depending on the level of understanding and the cultural background of the client, this informative process should be

conducted in an appropriate language with illustrations and imagery to enhance the spoken word.

More than one counseling session may be necessary and a summary of the session should be augmented with appropriate written materials, which the client can take home.

Confidential

Confidentiality must be maintained so that the client will be assured that his privacy is respected and kept. This will encourage full disclosure and create a trusting and beneficial relationship with the counsellor.

Non-directive

Counselling is not about telling clients what to do. It is about giving them accurate information to help them reach their own decisions. Directing them to take a particular course of action is often counter-productive. If the client later regrets the decision he or she will resent the directive and blame the counselor for it.

Where the client decides to ignore the directive the relationship with the counselor becomes threatened and may cease to exist. It is unethical for the counselor to subtly direct the client by providing unbalanced and biased information.

Supportive

The counselor should empathize with and support the decisions taken by the client. The counselor should help the client explore self, feelings, attitudes, and values in relation to the diagnosis. The client should feel secure enough to return and discuss possible consequences of his decision. The counsellor should refer the client to where he can obtain further information, social services or appropriate health care advice.

3.3.2. Objectives of Genetic Counseling

The counselor should;

1. ensure that the client has obtained an accurate diagnosis of his condition
2. ensure that the client is feeling well and comfortable and not at that moment in need of medical attention
3. ensure that the client knows what the session is about and how long it might last
4. take a family history pertaining to sickle cell or other inherited
5. haemoglobin disorder
6. establish the prior awareness, knowledge and perceptions of the client about SCD and sickle cell trait
7. tactfully but firmly dispel myths and misinformation held by the client
8. ensure that the client understands the inheritance of SCD and how it might affect him or his children depending on the Hb genotypes of their parents
9. ensure that the client understands the likely clinical course and treatment needs of patients with SCD
10. ensure that the client understands that carriers of the sickle cell gene are healthy persons who are unlikely to fall ill because of their status.
11. inform the client about existing facilities for health and social care and provide a referral where necessary
12. provide the client with a spectrum of careers that are compatible with living with SCD.
13. encourage affected clients and their parents by referring to successful role models living with SCD in the community summarise the session, allow for questions, handover educational materials and make appointment for a follow up session

Research, Research, Research

The importance of research is underestimated in Nigeria due largely perhaps to our under-development. There would be no progress without research into all aspects of sickle cell disorder. But for the research carried out in America, the present state of knowledge which has improved the quality of life and life expectancy in sickle cell anaemia would not have been possible. As the country with the largest burden and blessed with many bright scientists, we need the financial resources to devote to research that should convert sickle cell anaemia to a disorder like hypertension which is compatible with normal duration and quality of life.

Development of a National Policy on SCD

The SCFN has constituted a Nigerian Sickle Cell Expert Advisory Committee. The committee's first objective is to consider, initiate and revise policy and strategies appropriate to the management, prevention and control of sickle cell disorder in Nigeria. Other objectives and decisions can be found on the SCFN website www.sicklecellfoundation.com.

The First World Sickle Cell Day – 19 June, 2009

Following decades of advocacy, the United Nations in December 2008 resolved to recognise sickle cell anaemia as a public health problem and to mark the World Sickle Cell Day (WSCD) on June 19, every year, starting from 2009. The intention is that the WSCD will create awareness and draw attention of governments, donors and all stake holders every year, to what has been achieved and what needs to be done to address SCD in each of the affected countries.

3.4. Comprehensive Care in Sickle Cell Disease and Recommendations in Nigeria

Comprehensive care incorporates provision of holistic healthcare services including state-of-the-art diagnosis, standard therapies, preventive care, rehabilitative therapy, and other

ancillary services, by a team of specialists in a given location, with maximum accessibility for all patients. Comprehensive sickle cell centers are grossly lacking in Nigeria. Holistic care has been shown to provide better outcomes in sickle cell disease evidenced by significant reduction in mortality, hospitalizations, and blood transfusion rates among Nigerian patients [65]. As well, WHO recommends that in areas where hemoglobin disorders are common, special dedicated centers with a high degree of autonomy are required in appropriate numbers and locations, with a high degree of autonomy [66].

Treatment of SCD requires a multi-specialist team including professionals such as hematologist, pediatrician, orthopedic surgeons, plastic surgeons, ophthalmologists, nephrologist, specialist nurses, clinical psychologists, and social workers. Provision of comprehensive health centers is crucial to improving SCD disease outcomes in Nigeria. At such facilities, treatment should be tailored to individual patient's needs. At diagnosis, proper education regarding the nature of the disease, possible complications, and its prevention and treatment should be offered to the patient and parents. Regular health maintenance visits should be scheduled and patients should be counseled on the need for adherence [67].

Compliance on the part of the patient depends on having adequate information on the disease and confidence in the health professionals. Similarly, timely and regular medical education should be provided to health care professionals involved in management of SCD in order to improve their expertise and skills. Also, establishment of support groups among patients is encouraged.

Comprehensive care centers must possess facilities for outpatient care, day-case admissions (day hospital services), and hospitalizations on a 24-hour basis [68]. Patients should have direct access (including phone contact) to such centers and their physicians. For acute complications and emergencies, a quick triage is carried out and prompt therapy is instituted.

Standard protocols should be provided for management of specific complications, as well as general health maintenance. Scheduled review and strict adherence to protocols are advised.

Patients and parents should be counseled on avoidance of known precipitants of sickle cell crisis. Keeping a diary of pain episodes is helpful in identifying and avoiding triggers for pain crisis. Infections especially malaria have been reported as a major precipitant of sickle cell crisis among Nigerian patients. As such, vector control and chemoprophylaxis for malaria is recommended in all patients [69]. All forms of undue physical exertion or exhaustion should be discouraged.

Mothers should be regularly reminded about routine national vaccination schedule as well as vaccination against organisms to which SCD children and adults are particularly susceptible, especially encapsulated organisms. Adequate and regular hydration is important. At least 60–70 mL/kg of oral fluids or at least 1.5 L/m2 every 24hours is recommended [70, 66]. Hydration helps with haemo dilution, which reduces the propensity for sickling and vasoocclusion. Regular hydration also prevents dehydration which they are prone to due to impaired concentrating ability of the kidneys. Exercise caution with fluid administration especially in those with renal disease or severe anaemia.

Excessive fluids may precipitate pulmonary oedema and death. Moreover, physicians should administer, monitor, and encourage patient‘s compliance with routine medications at follow-up visits. Routine medications include prophylactic antimalarial [71] and folic acid. Others may include antioxidants, aspirin, and prophylactic antibiotic (oral penicillin from 2-3 months of life until at least age 5 in areas where pneumococcal infection is prevalent). Malaria has been described as one of the major precipitants of VOC for patients in Malaria endemic regions including Nigeria, hence the rationale behind continuous life-long chemoprophylaxis [71–73]. However, according to a local study, no significant benefit or

advantage was associated with routine chemoprophylaxis for malaria in SCD patients as both patients and controls had equal rates of asymptomatic parasitaemia and similar frequency of malarial attacks [74]. In Nigeria, the actual benefits of malaria chemoprophylaxis in SCD need to be clarified through further research. Early institution of broad spectrum antibiotics is recommended in febrile SCD patients [66]. Antibiotic use should be guided by local bacteriological profile and should be commenced after necessary bacterial cultures are taken. A switch to appropriate antibiotic is based on sensitivity pattern of the offending isolate especially if the fever is persistent (unresponsive to the former antibiotic).

Disease Modifying Therapies

Hydroxyurea Therapy. Currently, hydroxyurea (HU) is the only approved disease modifying drug in SCD used for selected patients above 24 months of age [75]. HU is a cytotoxic agent that has been mainly used in treatment of CML and other myeloproliferative disorders. Its usefulness in SCD is related to its ability to induce increased levels of fetal hemoglobin production in sickle cells thus mitigating tendencies for red cell sickling. The exact mechanism is not fully understood, but, as a ribonucleotide reductase inhibitor, it prevents formation of deoxyribonucleotides, causing Sphase arrest of all replicating cells, thereby inducing stress erythropoiesis, which favors increased production of fetal hemoglobin [24]. HU is also known to increase steady state hemoglobin levels and reduce leucocyte and platelet counts. Also, as a rheological agent, HU improves cell hydration, limits interaction of the sickle cells with the vascular endothelium, and acts as a nitric oxide donor [76]. HU is of benefit to patients with moderate to severe sickle cell disease. Indications for HU therapy include recurrent VOC (3 or more severe episodes requiring admission in the last 12 months), recurrent ACS (2 or more episodes in a lifetime), severe symptomatic anaemia, and recurrent priapism, alternative to transfusion to prevent new or recurrent stroke especially where transfusion is not feasible [75]. Usually, HU therapy is commenced at 10–15mg/kg once daily.

Baseline investigations prior to commencement of hydroxyurea should include FBC, reticulocyte count, % Hb F, electrolyte urea and creatinine level, liver function test, uric acid, and LDH levels. Full blood counts are monitored weekly for the first 4 weeks, fortnightly for the next 8 weeks, and thereafter monthly if the counts remain stable [77, 75]. Its dose is increased by 2.5 to 5mg/kg/day every 12 weeks (range of 4 weeks to 6 months) if absolute neutrophil count (ANC) >2000, Haemoglobin concentration >4.5 g/dL, and platelet count >80,000/ L [77, 76]. As marrow suppression occurs, HU is withheld to allow form arrow recovery and then restarted at a dose of 2.5mg/kg less than dose causing myelosuppression. This is known as the maximum tolerable dose [78, 76]. However, the ceiling dose for HU therapy is 35mg/kg [79]. Minimum time interval for evaluation of therapeutic efficacy is 6 to 9 months [80]. Hb F levels should be monitored. HbF level in excess of 20% significantly ameliorates the disease. Complications of HU include myelotoxicity, mouth ulceration, macrocytosis and megaloblastoid changes, nausea, skin toxicity rashes, and hyperpigmentation

[88, 194].

Haemopoietic Stem Cell Transplantation (HSCT). Suggested eligibility criteria for HSCT in SCD include the following [81, 82]: (A) age <17 years; (B) at least one of the following complications: brain infarct/ischaemia (MRI), secondary cognitive impairment with cerebral vasculopathy, severe and recurrent ACS, ≥3VOC per annum requiring hospitalization (>3 Hospitalizations for severe VOC in consecutive 3 to 4 years), moderate glomerular dysfunction, multiple epiphyseal aseptic necrosis, and grade I/II sickle chronic lung disease; (C) availability of HLA matched sibling donor. Exclusion criteria include donor with major haemoglobinopathy and one or more of the following: Karn of sky performance <70%, Portal fibrosis (moderate or severe), renal failure (GFR <30%), major intellectual impairment, stage III or IV chronic sickle lung disease, cardiomyopathy, or HIV infection. Older adults are considered less favorable candidates for HSCT due to the higher risk for severe organ

toxicities and greater susceptibility to severe graft versus host disease [83]. HSCT should be performed in centers experienced in transplant for sickle cell disease.

Future Therapies. Aside from Hydroxyurea, other promising drugs that have been shown to modulate Hb F production but are still under investigation/trials include decitabine, 5-azacytidine, and short chain fatty acids such as butyrates [84, 85]. Other novel therapies are also being investigated. Their therapeutic efficacy is designed based on their targets against specific pathophysiological processes in SCD such as the abnormal membrane cation transport systems, increased/stimulated red cell-endothelial adhesiveness, endothelial activation and vasospasm, cellular dehydration, prooxidant state, and hypercoagulability in SCD. Gardos channel blockers such as clotrimazole and its analog, Senicapoc (ICA 17043), have been shown to reduce red cell dehydration and abate haemolytic rate and are well tolerated in SCD patients [86, 87]. Administration of magnesium salts is also observed to reduce red cell dehydration by inhibiting the KCL co-transporter. It is reported that infusion of magnesium sulfate reduced the length of hospital stay in patients with VOC [88]. However, this is not yet an established practice. Similarly, anti-adhesive agents such as anti-P-selectin and heparin, as well as agents such as warfarin and aspirin for normalization of hypercoagulable state and Flocor for reduction of whole blood viscosity, and specific monoclonal antibodies for inhibition of red cell-endothelial adhesion are also being considered [84]. Inhalational nitric oxide and its precursor, L-arginine, are shown to be beneficial in acute vasoocclusive crisis and other ischaemic complications by increasing NO bioavailability [85, 88].

Theoretically, gene therapy offers a great hope of cure. However, effective vector for safe transfer and stable, erythroid specific expression of normal beta globin gene are still under investigation

[89, 90].

3.5. Implementing Comprehensive Health Care Management for Sickle Cell Disease in an African Setting

Sickle cell disease (SCD) is the commonest single gene disease in Africa.[91] About 40 countries in Africa and at least 23 countries of West and Central Africa carry the βs gene.[92] In Nigeria, 20-5% of the population are carriers of the genetic abnormality,[93] and about 3% of babies are born with the disease[94]. The basic pathology in SCD is the polymerization of haemoglobin following deoxygenation, a process that results in vaso-occlusion, which is the hallmark of the disease[95]. This process results in various forms of complications such as bone pains, abdominal pain, cerebrovascular accidents and priapism, which characterize the disease. In addition, children with SCD are prone to various forms of infections as a result of defective immunity arising from defective splenic functions,[96, 97] functional abnormalities of white cells [98] as well as abnormalities of the complement pathway.[99] The mortality rate in

SCD has remained high in Africa. [100-103]

potential for clinical cure in SCD. This modality of treatment is very expensive and thus, not available to a majority of children with SCD. Therefore, alternative treatment modalities which are effective in reducing the morbidity and mortality associated with SCD as well as improving the quality of life in SCD are highly desirable.

The management of SCD continues to pose a challenge in Africa. Not only does the populace have poor knowledge of the disease,[104] the healthcare providers may also not be familiar with the current concepts in the management of the disease. The management of SCD in most health facilities in Africa largely addresses the complications of SCD rather than the disease itself hence, morbidity and mortality in SCD has remained high. In some developed countries, comprehensive health care was adopted as the strategy for the management of SCD and this has resulted in the reduction of mortality from 16-30% to less than 1% in that setting.[105-108] This method of care, as practised in the developed countries, requires the

involvement of several professionals such as the genetic counsellor, paediatric haematologist and the social worker among others. This multi-disciplinary approach offers a holistic form of care to children with SCD and ideally should be the model of care. In Africa and other developing countries where SCD is prevalent, the form of care described above is not within reach due to dearth of most of the required health professionals.

Therefore, there is a need to adapt this form of care for the low-resource settings in Africa where SCD is prevalent. This model of care has been successfully adopted in Cotonou, the Republic of Benin. This resulted in a drastic reduction of morbidity and mortality associated with SCD in the country. In recognition of this adaptation, the World Health Organization, at the meeting of the Regional Committee for Africa in June 2010, proposed that by 2020, half of the 23-Member States with high prevalence of SCD should have developed and commenced the implementation of a clearly designed National Sickle Cell Control programme within the context of a National Health Strategic Plan. It is also expected that 25% of the countries in the African Region should have adopted the concept of comprehensive health care management by the year 2020. [109] This strategy was adopted by the Sickle Cell Centre at the Republic of Benin in 1993 [110] and the University of Benin, Benin City, Nigeria in 2003. In Cotonou, the Republic of Benin where this strategy of care is being utilized, the mortality of SCD dropped to 15.5/ 10000, a value ten times less than the overall Under-Five mortality rate in that country. [111] In Italy where this strategy was also adopted, the morbidities and mortality associated with SCD have reduced. [112]

In the last 13 years, the Paediatric Haematology Unit of the University of Benin Teaching Hospital has managed 547 children with sickle cell anaemia (SCA) using the new protocol. Twenty children with SCA died over this period; the majority of the deaths occurred among

[103] children referred from other facilities on account of complications of SCA. Seven children in the new protocol died; two from acute chest syndrome, one from liver failure, one

from sequestration crisis (died in a private facility) and the rest from sepsis. In the last five years, all the deaths among children in the programme occurred at the Children Emergency Room before the Paediatric Haematology Unit could have an input in their care. The admission rate for sickle cell crises has reduced to an average of one in about two years with some children being crises-free for five years. The transfusion rate has also reduced drastically to less than 2% and the mortality rate amongst our cohort is 1.3% (7/547). Using the Body Mass Index (BMI)-Z scores in a recent nutritional survey of 187 children with SCD, the nutritional status of the children in this programme was similar to that of their peers, with a reduction in the prevalence rate of severe malnutrition to less than 18%. Indeed, 75.9% had normal nutritional status while 7% were overweight. With these achievements, it became necessary to share this method of care so healthcare providers in other parts of the country and beyond, can adopt same and more children with SCD may benefit from its numerous advantages.

3.6. Management of Sickle Cell Disease

It is important to stress that the management of SCD is not synonymous with the management of the complications of the disease. Patients with SCD only have chronic anaemia as a constant feature of the disease. While chronic anaemia is the only constant feature of SCD, other morbidities associated with the disease are actually complications of the disease. The aim of managing SCD includes the prevention and early recognition of the complications of the disease. These complications are, usually, the causes of death in SCD.

The concept of Comprehensive Health Care management of SCD is hinged on the following constituent services which are provided by the clinician during clinic visit:[113]

- Parental counseling and education.
- Education on the need for adequate nutrition.

- Education of the need for adequate hydration.
- Early identification of fever and its urgent treatment
- Early identification of a large spleen.
- Use of prophylactic medications like Penicillin V, anti-malarial drugs, Folic acid and Vitamin C.
- Immunization against infections which are common to children with SCD such as *Salmonella*, *Hemophilus influenza* type b, Pneumococcus and Hepatitis B virus infections.
- Need for regular hospital follow-up.

Parental Counseling and Education

The knowledge of how sickle cell disease is acquired has remained very poor amongst Africans despite the high prevalence of the disease in this area.[77] Therefore, marriage between carrier individuals persists without inhibition.[77] Usually, it is the presence of the disease in an affected child that brings the carrier status of the parents to fore. Only very few intending couples practice pre-marital screening for SCD particularly where churches insist on screening before marriages are contracted.

In situations where couples co-habit and where marriages are contracted according to traditional practices, screening for SCD is usually not done. In counseling couples with a child who has sickle cell disease, they are taught how the child acquired the disease and the effects of the disease on the child are explained to them to enhance a thorough understanding of the pathophysiology of the disease. The aspect of education is very important in an African setting where mothers are usually blamed for the production of ―bad children‖ or witchcraft when families experience childhood deaths attributed to SCD. In addition, the knowledge of the patho-physiology of the disease helps parents to manage the child with SCD better and

comply with the protocol of care.[114]The care of the children enrolled into this programme is accomplished in the homes of the affected families rather than in the hospitals, hence, the need for parental education.

Adequate Nutrition

Malnutrition is a common problem among African children. A lot of African children are stunted due to poor nutritional intake. In Nigeria, 41% of the general populations of children are stunted. [115] It is this same setting that has the bulk of children with SCD. Specifically, SCD poses a high nutritional burden on the affected children.

The disease is associated with a high metabolic rate because of the bone marrow hyperactivity.[116, 117] The life span of the sickled red cell is 10-20 days instead of the 100-120 days for normal haemoglobin. This places a high nutritional demand on the child with SCD. In addition, a child with SCD also requires the same nutrients for his growth and development. This implies that children with SCD on diet similar to that of healthy children are at risk of failure to thrive in early childhood and stunted grow with delayed secondary sexual characteristics later in life. These children are usually thin with asthenic build. With adequate nutrition, children with SCD are able to achieve normal growth and development as their peers with normal haemoglobin. The stigmata of the disease such as prognatism, skull bossing and long thin extremities have been observed to disappear with adequate nutrition. Therefore, caregivers are advised to feed their children frequently with as much as six to eight meals daily using nutritious diets. Non-nutritious meals or snacks should be discouraged and the parents are advised to ensure children go to school with food instead of snacks.

Adequate Hydration

Vaso-occlusive events occur frequently among children with SCD because the sickled red cells cause hyperviscosity of the blood which results in sluggish blood flow. Dehydration worsens this hyperviscosity and the latter reduces the flow velocity and thus, increases the delay time that is critical in polymer formation.[118] Polymers are responsible for vaso-occlusion which is the hallmark of SCD. Most of the morbidities and mortality which characterize SCD result from multiple organ damage following recurrent vaso-occlusive events. Therefore, the prevention of vaso-occlusive events is important in ensuring not only the wellbeing of the SCD child but also the prevention of chronic organ damage and early death. Children with SCD are advised to drink 2-3L /m2 of water daily. The child is advised to go about with water in order to facilitate adequate water intake and good hydration. Adequate hydration also enhances the urinary excretion of bilirubin resulting from the chronic haemolytic state in SCD. Children with SCD in our programme remain crisis-free for as long as two to five years following adequate water intake. Hydroxyurea, an anti-sickling drug, is not routinely used for the children with SCD enrolled in the programme hence the use of this medication may not explain the observed improvements among our cohorts.

Early identification and treatment of fever

Fever is regarded as an emergency in a child with SCD as fever may be a flag sign for infections. Infections are common causes of morbidities and mortality among children with SCD since their immune functions are usually poor.[119] Caregivers should be taught how to detect fever in children using a thermometer or by feel and report to the hospital once the child with SCD is noticed to the febrile. In most parts of Africa, this advice is very pertinent as most caregivers patronize drug vendors for medications when their children are ill. This

contributes to delay in seeking appropriate care for the child with SCD. It is important to note that pneumococcal infections have a doubling rate of 30 minutes.[120]

Early identification of a large spleen

Splenic function among children with SCD is defective as early as the sixth month of life. However, the most life threatening splenic event in SCD is the sequestration crisis.[121]This event results in the pooling of sickled red cells in the spleen with a resultant severe anaemia and severe hypovolaemia or shock. This event can be fatal if it is not promptly identified and appropriately managed with blood transfusion.[121] Caregivers should be taught how to identify a large spleen in a child. In Africa, identifying a large spleen by caregivers is important as its presence has been associated with anaemia and frequent illness. In traditional parlance, scarification marks are made over the abdomen to ―treat the spleen‖. A large number of children with SCD in Nigeria have these scarification marks on their anterior abdominal wall.

The caregivers should be taught how to detect a large spleen in their children during bath. The caregivers are advised to take the child to the hospital if they notice an enlarged spleen for thorough evaluation especially to check the packed cell volume.

Use of prophylactic medications

Drugs routinely given to children with SCD include Folic acid 5 mg daily, Vitamin C and malaria prophylaxis. Folic acid therapy prevents megaloblastic crises as a result of high red cell turnover, Vitamin C functions as anti-oxidant because of the oxidant stress imposed by the red cell hemolysis[122] while malarial prophylaxis is important for children in malaria-endemic region.[123] Malaria not only worsens the anaemia but is also a cause of frequent morbidity. Penicillin V tablet is administered for the prevention of pneumococcal

infections.[124, 125] For children less than three years, Penicillin V 125 000 IU is administered twice daily while the dose is doubled for children aged between three and five years. The drug is used along with the pneumococcal vaccine.

Immunization for the child with SCD

Infection is the commonest cause of death in SCD children, especially infections due to encapsulated organisms [119, 120, 124, 126-128]. Therefore, vaccinations against *Haemophilus influenza* type b,[129] pneumoccocus[130, 131] and Salmonella are essential in the care of children with SCD. However, the routine immunization schedules in most African countries do not include these vaccines. Nigeria, for example, has just implemented the use of the *Haemophilus influenza* type b and pneumococcal vaccines for routine immunization. Salmonella vaccine is not included in the routine immunization schedule for children in Nigeria. These bacterial organisms have been proven to be the major causes of infections in children with SCD, hence, there is an urgent need to vaccinate these immune-compromised children to prevent morbidities and mortality associated with infections.

These vaccines are routinely used for the children with SCD enrolled into the programme at the University of Benin Teaching Hospital, Benin City. Hepatitis B vaccine was only introduced 12 years ago in Nigeria, hence, children with SCD who are older than 12 years must have missed the vaccines. Therefore, children who are older than 12 years and those aged less than 12 years but who were not vaccinated should be tested for Hepatitis B virus infection and the vaccine should be administered if they are sero-negative.

Regular Hospital follow-up

For a disease that has lifelong implications, there is a need for regular hospital follow up. In Africa, most patients with SCD do not attend follow-up care in the hospital setting. Children

are only brought for care when they have crises. This attitude may be is detrimental to the health of children with SCD because regular follow-up hospital visits may ensure adequate growth and development through growth monitoring and frequent education. This allows for early identification and prompt management of complications. This allows the child with SCD to maintain good health.

3.7. Sickle Cell Disease: The Role of Self-Care Management

Sickle cell disease (SCD) is a group of genetic disorders that is characterized by the development of abnormal hemoglobin (Hgb S), abnormal red blood cells, and the resultant complications [132]. This disease has a high prevalence in Africa, the United States (U.S.), the Caribbean, Central and South America, Saudi Arabia, India, and the Mediterranean [133]. Approximately 90,000 to 100,000 Americans are affected by SCD, making it the most common genetic disorder. In the U.S., this disease primarily affects African Americans. Within this group, there is an incidence of one in 500 births and the sickle cell trait occurs in about one in 12. Sickle cell disease is an autosomal recessive disorder, and males and females are affected equally [134]. Diagnosis of SCD is typically made at birth during newborn screening. Disease management focuses on pain, hydration, and preventing infections and other complications that result in vaso-occlusive crises [136]. The annual estimated costs of medical care for SCD patients total over $1.1 billion; 80.5% of the costs are attributed to inpatient hospital care, 3.2% to emergency department (ED) use, 0.9% to physician visits, and 3.6% to prescription drug use [135]. These facts altogether suggest that SCD has a high public health importance as it places a burden on not only the community of the affected population but also the health care infrastructure.

Abnormal, c-shaped or sickle-shaped, hemoglobin molecules impede circulation through the blood vessels [132]. The cells adhere to each other and to the walls of blood the vessels,

blocking blood flow, resulting in decreased circulation to body tissues, decreased oxygen capacity of the red blood cells, organ damage, and pain (vaso-occlusive pain crises). These abnormal red blood cells have short lives and hemolyze frequently, leaving the affected individual in a constant deficiency of red blood cells that results in anemia. Four major types of SCD exist with sickle cell anemia being the most common [137, 138]. Major complications of SCD include vaso-occlusive pain crises, acute chest syndrome, acute splenic sequestration, pneumococcal disease, and cardiovascular accidents [139]. These complications, along with chronic renal failure and pulmonary disease, are associated with risk of early death in this patient population [140]. The only cure for SCD is bone marrow transplantation, but there are few potential donors available, and the procedure can have fatal side effects [141]. Hematopoietic stem cell transplantation has shown promising outcomes, particularly when donors are human leukocyte antigen (HLA) matched siblings; however, older age (transplantation has optimal outcomes during childhood) and advanced disease are associated with poor transplant outcomes [142].

The sickling of the cells causes impaired blood circulation which results in pain. This is the most common complication of sickle cell disease. It can begin as early as infancy and can happen unpredictably throughout life [138, 143]. Sites of pain often include the long bones, joints, the back, the abdomen, and the chest [144]. The pain may be mild, moderate, or severe, and has been described as sharp, throbbing, stabbing, deep, achy, lacerating, or shooting[145]. These pain episodes are the most common reason for hospital visits [146, 147]. On average, adults with SCD visit emergency departments (ED) over 197,000 times annually; 67% of individuals report pain or unspecified pain as the reason for the ED visit and 29% result in hospital admissions [147]. Approximately 90% of hospital admissions are for the treatment of acute pain[146]. The frequency of pain crises is indicative of disease severity as they were found to be associated with premature death in individuals over 20 years of age[134]. Pain

episodes are most frequent between ages 19 and 39 [143, 147]. On average, adults have at least one vaso-occlusive pain episode annually[148]. Utilization of the acute care system, however, is highest (3.61 instances per patient per year) for 18 to 30 year olds and in this group, the rate of re-hospitalization within 30 days is 33.4% [149]. Further, patients ages 18 to 30 are more likely to visit multiple hospitals for acute care; this may result in poorly coordinated and poor quality of care [150]. A typical crisis lasts for approximately 10 days, with women reporting longer episodes than men. Physical exertion, extreme temperatures, and stress were noted as aggravating factors [148]. A typical medication regimen for an individual with frequent pain episodes includes hydroxyurea therapy, analgesics, and possible red blood cell transfusion therapy [134].

Prevention of pain crises is essential to the health maintenance of individuals suffering from SCD. Management of this disorder primarily occurs at home and the focus is on these crises as they often result in ED visits and hospitalizations [140]. General recommendations from the Centers for Disease Control and Prevention [151] are that these individuals should receive regular checkups at least annually, stay hydrated, eat a healthy diet, get enough rest, avoid temperature extremes, and prevent infections. Other strategies for managing pain at home include medications, massage therapy, hot showers, distraction, and relaxation

[152, 140]. The

use of massage and relaxation/meditation was noted to increase with higher pain intensity[153]. Analgesic therapy, both opioids and non-opioids, is commonly used to treat acute pain in the home setting. Non-opioids most frequently include non-steroidal anti-inflammatory drugs [154]. Ibuprofen, acetaminophen, and an acetaminophen codeine mixture are used often [155]. Opioids may include morphine, hydromorphone, oxycodone, methadone, codeine, or tramadol [154]. Exchange transfusions and hydroxyurea therapy are also used as preventive strategies to reduce levels of Hgb S and have been helpful in decreasing pain, but these do not completely prevent crises [146, 134]. Some barriers to providing effective pain relief in the home

were identified as limited knowledge of coping strategies and inadequate knowledge about the use of analgesics to control pain [155].

A review of literature related to sickle cell disease (SCD) is presented here. The information is organized according to critical concepts associated with management of the disease. These concepts include self-efficacy, social support, socio-demographic variables, and self-care management. Principles of chronic disease management related to these variables are also included here.

Self-Efficacy

Sickle cell disease self-efficacy was evaluated in this study; however, the term is not widely published in the literature. In the absence of this information and for the purposes of this study, the principles of self-efficacy were applied to SCD self-efficacy. Self-efficacy links self-perceptions with individual actions, is a mediator of health behaviors, and is considered crucial to chronic disease management [156]. In studies that evaluated chronic conditions, higher levels of self-efficacy were linked to decreased pain severity and fewer self-reported symptoms [158] as well as lower levels of depression, stress, and anxiety [157]. Lower levels of self-efficacy were correlated with more symptoms, higher pain severity, and frequent physician visits. In addition, higher levels of self-efficacy were associated with increased use of adaptive coping mechanisms, decreased anxiety and stress, as well as increased adherence to medical regimens [157]. In a study involving cancer patients experiencing pain, lower self-efficacy for pursuing medical information was associated with greater barriers to pain management. An inverse relationship between distress and self-efficacy for coping was also noted [159]. In SCD, self-efficacy was negatively correlated with the number of crises per year [459]. It was also noted that self-efficacy beliefs were inversely related to symptomatology and health care utilization, and that these beliefs may be predictive of future changes in SCD symptomatology

[158, 157].

Social Support

The quality and availability of social support may influence the health outcomes of individuals with chronic illness and may positively affect self-care behaviors[160]. Types of social support include emotional, instrumental, informational, and appraisal support. Support may be obtained from family, friends, work, and the community [161]. Patients' perceptions of satisfaction with this support are important as they have been linked with adherence to medication self-monitoring in individuals with chronic conditions. In chronic disease states, social support from various sources is important. Family support may decrease depression and increase compliance, while support from healthcare providers might increase satisfaction with the healthcare system and decrease perceived discrimination[162]. Children with family social support were found to have better disease management behaviors [163]. Healthcare provider support improves long-term self-care management and the ability to manage chronic conditions [164]. In addition, social support was noted to be helpful in following treatment plans [165, 166, 164].

Socio-demographic Variables

Socio-demographic variables are strong predictors of health related quality of life in individuals with SCD. These variables include age, sex, income, education, employment, and marital status [167]. Panepinto and Bonner [168] note that health related quality of life (HRQL) in adults with SCD is significantly impaired and may be worse than in other chronic diseases. Variables affecting HRQL in SCD include age, gender, family demographics (education and income), disease-related symptoms, rural versus urban residence, and employment. Age and socio-economic status, for example, negatively affect HRQL. African Americans with SCD were more likely to be less educated, have lower income, and were more likely to be unemployed or disabled when compared to African Americans without SCD [169].

Socio-demographic variables appear to significantly affect patient outcomes. Researchers typically operationalize these variables using age, income, education, and employment or occupation. Given that minorities are typically disadvantaged in the majority of these areas and are primarily affected by SCD, these variables should be accounted for when attempting to improve health outcomes. Further research is needed regarding assessment of and strategies for dealing with socio-demographic problems experienced by SCD patients [15].

Self-care Management

The concepts of self-care and self-care management (often used interchangeably with self-management) have been increasingly studied over the years. This is due in part to the rising prevalence of chronic diseases and higher rates of healthcare utilization. In light of this finding, self-care management is important in improving health outcomes, enhancing quality of life, and decreasing healthcare costs [170]. Self-care refers to learned, routine, positive, and practical activities performed by individuals to maintain life, health, and well-being. Self-management is the ability of an individual with a chronic disease, to participate in a daily, self-motivated, collaborative (conducted with family, social, and healthcare provider support) process to manage symptoms [171]. This process involves the domains of focusing on illness needs, activating resources, and living with a chronic illness. In chronic conditions, an individual‘s ability to perform behaviors that will alleviate the pain experience is instrumental in adapting to pain long-term [172]. Common definitions of self-management, however, are somewhat broad and cover not only a variety of behaviors but a variety of conditions, thus leading to confusion regarding its application to research and practice [173]. Given this finding, it is important to understand self-care management and self-care behaviors in the context of SCD. Overall self-care management strategies specific to SCD have been identified. In a study assessing strategies used by adults with SCD to manage their illness, themes included self-awareness, emotional support, career selection and success factors, nutrition, advocacy,

knowledge, physical activity, and complementary and alternative medicine [174]. In middle-aged and older adults, identified self-care recommendations were physiologic (warmth, hydration, rest, good food, and avoiding drinking, smoking, and using drugs), psychological (knowledge and understanding of the disease, listening to and learning about the body, prayer, and social support), and provider-related (knowledgeable health care providers and following providers‘ orders) [175]. There are many benefits of self-care management; therefore, it is important to understand which factors affect self-care management. Further, an evaluation of self-care management in the young adult age-group is necessary.

The Relationship among Concepts

According to the theory of self-care management for SCD, vulnerability variables (socio-demographics and health needs) negatively affect health outcomes (health status) and self-care management resources (including self-efficacy, social support, self-care ability, and self-care actions) [167]. Self-care management and coping may be correlated with self-efficacy in that it reduces the effects of stressors and facilitates positive health behaviors[157]. In a study conducted with diabetics, another population requiring self-care, self-efficacy was significantly correlated with positive outcome expectations in the prediction of diabetes self-management and hemoglobin A1C [176]. When positive expectations were high, high self-efficacy was related to better self-management. Self-efficacy, in conjunction with self-care ability, assertiveness, and social support, may assist with the management of daily activities necessary for coping in SCD [167]. Socio-demographic variables affect functional ability and health related quality of life [168]; therefore, these variables likely affect perceived self-efficacy, the availability of needed resources and social support, and participation in self-care management.

In the majority of the studies reviewed, data was obtained mainly or only using self-report. Researchers often consider this a study limitation as participants responses can be variable

and unreliable, even on similar measures used within a given study. There are benefits, however, to using this method of data collection. Direct reports from participants about their experiences provide rich information. This adds depth to and enhances the understanding of the phenomenon of interest; qualitative data adds meaning to quantitative data. Also, for concepts such as pain and perceived self-efficacy, subjective report is the only way to obtain accurate information, making it the gold standard.

3.7.1. Conceptual Framework

In regard to sickle cell disease and the number of hospital visits for crises, self-care management, self-efficacy, social support, and socio-demographic variables appear to be the most significant constructs discussed in the literature. To address the relationships among these variables, a Model of Health Functioning was developed by the author to explain the mechanisms of self-care management as it relates to sickle cell disease; it outlines the predictors, mediators, and outcome. The model also provides the framework for conducting data analysis. According to the model, SCD self-efficacy, social support, and socio-demographic variables are related to self-care management (perceived self-care ability and self-care action)[160, 157, 156, 167, 177]. Sickle cell disease self-efficacy, social support, and socio-demographic variables (age, gender, level of education, employment status, income, and living situation) are associated with self-care management which in turn is associated with the number of hospital visits for crises in this population. There are also direct relationships among SCD self-efficacy, social support, and socio-demographic variables in understanding hospital visits for crises. In addition, self-care management plays a mediating role between the predictors and the outcome.

In summary, to decrease the number of hospital visits for crises, prevention is imperative. Self-efficacy and self-care in the home setting may be integral to pain self-management and thus crisis prevention [157]. Self-care is correlated with a number of variables, but it is unclear

which one has the greatest correlation with decreased pain severity for SCD patients. The mechanism of this relationship also needs to be clearly defined. Studies show a positive correlation between self-efficacy, social support, and self-care activities in SCD patients [156], but there is still some question as to which self-care activities are most effective and which variables affect self-efficacy and social support.

In addition, it would be beneficial to further evaluate socio-demographic variables in relation to SCD. Variables that play a role in the number of hospital visits for crises need to be identified and the relationship quantified before interventions can be formulated.

3.8. Community Health Workers as Support for Sickle Cell Care

Sickle cell disease (SCD) is a set of genetic hemoglobinopathies characterized by hemolytic anemia, severe pain, and multi-organ compromise. As a result of dramatic improvements in care and longevity,[178–180] SCD increasingly has become a lifespan disease, from childhood to adulthood.[181] However, approaches to managing SCD encounter problems at multiple levels that challenge both the patient and healthcare system. The purpose of this paper is to summarize the potential for community health workers (CHW) in the care of people with SCD, offer practical guidance for the development of new CHW programs for SCD through current examples, and highlight evaluation and policy opportunities.

Sickle Cell Care Highlights Gaps in a Socioecologic System

Though considered a rare disease by U.S. standards, SCD has come to be regarded a significant target for U.S. healthcare reform efforts, such as Healthy People 2020.[182] Much recent attention to SCD has arisen from the high morbidity, healthcare utilization, and cost associated with SCD, both for individuals and healthcare systems. [183–193]

therapies have led to improved life expectancy, many outcome goals remain unmet, not only because of the biological burden of end-organ failure or acute complications but also because

of the complex burden of SCD patients' interaction with the socioecologic system.[194–196] This system emphasizes the inter-relationship among individual, interpersonal, organizational, community, and public health factors in relation to health behavior change.[197] Like other chronic conditions, SCD is influenced at all of these levels by varying factors, and by interactions between these factors. Interaction between disease manifestations and health behavior is only one part of the challenges to the individual within the socioecologic system.

Regardless of management or adherence, pain and other disease manifestations can be acute or chronic, incapacitating, and unpredictable[198–201]. Thus, patients and caregivers must learn disease self-management, how to access healthcare systems, and how to make healthcare decisions. Unfortunately, mismatches between the needs of the SCD population are often entwined with health disparities associated with race/ethnicity in the U.S.[194–196], including those at primary[202] and specialty care levels.[203] Contrary to best practices for care of chronic health conditions,[204] only a fraction of Americans with SCD receive regular medical care for scheduled preventative care and evaluation. SCD care is often delivered in resource-intense medical settings such as emergency departments and in-patient hospital stays, especially after childhood.[205, 206] Even hematologists might not prescribe hydroxyurea,[207– 209] the only U.S. Food and Drug Administration–approved disease-modifying drug for SCD.[208,209]

At the community level, impediments to helping people with SCD maximize function at school and work contribute to high unemployment and disability.[210, 211] Social isolation may be compounded by cognitive impairment and stigma of SCD.[212, 213] In total, these multilevel issues lead to high variability in patient satisfaction, quality of life, and adherence to

prescribed medical regimens and appointments. These multilevel barriers should be addressed to improve care and outcomes for people with SCD.[214]

Community Health Workers

Intervention by CHWs can target barriers to improved health and well-being for people with SCD on multiple levels. CHW programs aim to address the needs of underserved populations by providing trained community members as educators, supporters, and advocates who can effectively reach their neighbors.[180–217] In health care, CHWs are non-clinicians who work with medically and socially complex individuals from underserved communities to help bridge to medical services to improve patient healthcare engagement, self-management, treatment plan adherence, and health outcomes[218]. CHWs are called varying terms, including lay health worker, outreach worker, health advocate, promotora de salud, patient navigator, and guide. Such terms convey the concept that CHWs can effectively connect to multiple levels of a system. CHWs often share responsibility for patient education, support, and social services with clinic staff, and they have been successful in engaging and sustaining comprehensive disease care.[214–216]

Seven core roles in improving patients‘ health can be served by CHWs, who provide:

1. cultural mediation;
2. informal counseling and social support;
3. culturally appropriate health education;
4. advocacy for individual and community needs;
5. assurance that people receive needed medical and social services for which they are entitled;
6. direct social and supportive services; and
7. support for building individual and community capacity.[216]

The contributions these core CHW roles make to behavior change are supported by social cognitive theory.[218, 219] This construct states that behavior is shaped and maintained by consequences, particularly by immediate feedback from both objective sources (such as blood results) and an individual's social network (beliefs and traditions of family and friends).[220] CHWs could also help patients move between stages of the transtheoretic model of behavior change for self-care and chronic disease self-management. In this model, processes for adoption of positive behaviors are linked to enhanced decision making and self-efficacy.[221, 222]

Examples of the multilevel functions of CHWs can include the level of coaching the individual with SCD in adaptive behavior[223]. At the healthcare system level, CHWs can be integrated into the clinical setting as Patient Navigators to assist the patient with access to medical care. CHW integration with the medical team is highly effective in adult diabetes[224– 226]; pairing CHWs with nurses improves outcomes more so than nurses working alone.[227] On the community level, CHWs can engage, support, and advocate for families through schools and social service agencies.

The evidence for CHWs to impact these multiple levels in other disease areas is strong, as reflected in several systematic reviews.[228–230] CHW programs have existed for more than a century in developing countries for infectious disease management[231] and maternal child health.[232–234] Among the adult chronic diseases, CHWs have been shown to improve health outcomes for asthma, hypertension, heart disease, diabetes, and HIV, as well as improve disease self-management, disease prevention, pregnancy outcomes, and healthcare utilization.[227,235–238] For children with chronic disease, evidence of the benefit of CHWs is particularly strong for asthma.[219,239–241] In addition, studies indicate that CHW interventions are cost effective, especially when taking a longitudinal view of the return on investment.[229,

[242–246] CHW interventions have specifically been shown to lower hospital readmissions.[247, 249] Because of the critical role CHWs play in health and the emerging needs of the workforce, the American Public Health Association established a specific section on CHWs.[250]

The diversity of CHW roles and terminology shown in the SCD projects is similar to the published literature on CHWs in other fields. Many CHWs have a role that can be called Patient Navigator, whereas other CHWs focus on home visiting and improving access to community resources. Generally, CHWs are viewed as non-clinical professionals, as opposed to a healthcare professional like a nurse, social worker, or physician providing home visits or community outreach. CHWs may also participate in tasks such as coordinating medical care and providing assistance for diverse needs such as health insurance enrollment, immigration, or housing. These types of efforts serve to improve not just the medical but also the social health of the individual.

Social and cultural alignment of CHWs with their communities is a major aspect of effective intervention. They can be community-based or hospital-based, but regardless of their point of patient interaction, they are most effective when they share common cultural, social, and linguistic aspects with the clients they serve. For comprehensive, culturally appropriate SCD care, CHWs can address many of the barriers to access, communication, support, and disease self-management while reinforcing healthy behavior. Culturally aligned CHWs are an accepted mode of outreach by community members, sometimes in the context of community-based organizations for improving the health of underserved communities.[244, 250, 251]

3.8.1. Reported Use of Community Health Workers in Sickle Cell Disease

The use of CHWs for SCD has been limited. In the U.S., CHW programs have focused on common diseases. By contrast, based on population prevalence, SCD is considered rare. In

the global public health sphere, WHO recognized SCD as a priority topic in 2006,[252] but many countries with the highest incidence of SCD still struggle to balance prioritization of non-communicable and communicable diseases in low-resource settings. To the best knowledge of the authors, CHW interventions dedicated only to SCD have been published only in three reports. In Ghana, home visits by CHWs help families cope with new diagnosis of SCD in a newborn screening program.[253] In a peer mentoring pilot program at St. Christopher's Hospital for Children, older adult volunteers who raised a child with SCD advise new families on medical and child-rearing practices.[254] In St. Louis, CHW home visitors promote early cognitive development, promote clinic attendance, and medication compliance.[255] A recent systematic review of CHW interventions for children with chronic health conditions listed SCD as appropriate for CHW intervention and research.[232]

3.8.2. Conceptual Bases and Potential Roles for Community Health Workers in Sickle Cell Disease

Social cognitive theory can guide CHW interventions at the individual and family levels.[220, 256] Behavior is shaped and maintained by consequences and an individual's social network (beliefs and resilience of family and friends).[257, 258] For example, repeated clinic visits may not be perceived by patients and families as addressing pressing psychosocial needs. Poor alignment with provider–patient needs may lead to patient frustration and perceived gaps in addressing needs. By contrast, in home visits, CHWs can check for potential triggers of acute problems: disorganized medications, inadequate hydration or nutrition, physical barriers, family stress, and household chaos. The SCD CHW must support the positive behaviors and social networks using self-management skills.

Interventions by CHWs for SCD can be viewed through the socioecologic model. At the individual level, lifestyle, medication adherence, and coping are important issues for

individuals with SCD, and CHW interventions have demonstrated impact in these areas for other chronic diseases. Aspects of self-management for SCD can be promoted by coaching in community-based mentoring programs, peer groups, and others. At the family level, multiple social determinants of health can benefit from CHW programs, including maternal health and economic self-sufficiency[234]. CHWs may also be able to help the family understand reproductive choices relevant to the sickle gene, which can be passed along to future children of the patient with SCD and by any family members who have sickle cell trait. At the healthcare system level, CHWs can help the SCD patient navigate multiple appointments, provide reminders of appointments, and organize transportation. At the level of schools or jobs, CHWs can coach the SCD patient in requesting accommodations for their disease, using the appropriate resources under existing rules for people with disabilities.

Application of limited resources may require CHW services for SCD to draw upon other CHW programs. For example, home visitor programs for early childhood can benefit academic readiness for children with SCD[234]. SCD early childhood home visitor programs might effectively address the neurocognitive impairment[259, 260] and frequent school absences associated with SCD[261]. Improving control of asthma as a comorbidity of SCD would address the increased risk of SCD complications in preschool children with asthma as a high-risk group,[262] and many CHW programs have shown benefits for asthma.[221, 256] In the socialized medical system of Brazil, one report describes the network of neighborhood CHWs who assist everybody with healthcare needs, and their main role in SCD care is ensuring that patients go to subspecialty care at the regional center for blood diseases.[263]

3.8.3. Evaluating the Impact of Community Health Workers on Sickle Cell Disease

Impact of CHW and other workforce interventions should include improvement in four primary domains:

1. outcomes related to health and quality of life for the patient and family;
2. access and utilization of standardized preventative care and acute care services;
3. patient satisfaction with communication and care; and
4. long-term medical, psychological, and social metrics of well-being.[264]

Metrics often used for quality of SCD care involve utilization of acute care services, for example, number of acute care visits, length of hospital stay, or hospital readmissions within 30 days. However, cost considerations may inadvertently push CHWs to drive these cost-based, organization-focused outcomes, potentially placing CHWs in conflict with their patients.[213, 265] Accordingly, the benefit of interventions to educate patients or coordinate care with the goal of reducing acute care utilization might require alternate approaches.

Adherence to preventive care, including prescribed medication use and attending scheduled visits, is recognized as a key measure of quality care.[266] Some of these adherence measures could be well-suited efficacy measures for SCD. CHW contact with the family shortly before a well-child visit has resulted in fewer missed appointments, fewer sick and acute visits, better understanding of anticipatory guidance,[267] and transition readiness.[268]

At the interpersonal level, CHWs operate as a vehicle for delivering educational content and encouraging behavior change. Pain-related poor quality of life in people with SCD[269] and social isolation are frequent concerns.[210, 211] The Patient Reported Outcomes Measurement Information System (PROMIS) question bank was developed by NIH to assess quality of life across multiple populations (www.nihpromis.org). The Adult Sickle Cell Quality of Life Measurement Information System (ASQ-ME) question bank was developed specifically for quality of life in SCD, using PROMIS methodology.[270, 271] Quality of life in SCD has been reported using the SF-[211. 272, 273] Unfortunately, behavioral change promoted by CHW does

not guarantee quiescent symptoms of SCD, even with complete adherence to care plans.[198–201]

Finally, intervention process measures are useful for defining essential and satisfactory elements of CHW interventions[274]. Quality assurance measures and quality improvement processes are also important to assure ongoing intervention.

Community Health Worker Certification and Training

As with other powerful tools for improved health, critical selection of CHW personnel, training, goal setting, reporting, and other structure are crucial for success.[275, 276] CHWs must be selected for excellent interpersonal skills, experience, empathy, and problem solving. Training curricula could be suitable for adapting flexibly to trainees of different backgrounds. CHW training generally features role playing and problem solving, experiential training, and meetings between CHW trainees and SCD providers.

Several training components have been identified by projects with SCD CHWs:

1. basic training for CHWs for any disease condition;
2. specific training material on SCD;
3. experiential learning by shadowing in the SCD clinic; and
4. continuing education.

Experienced trainers of CHWs emphasize person-to-person skills, such as respect, patient centeredness, and communication. Practice and application of new knowledge are essential. Role play, teach backs, and other adult education strategies could help prepare CHWs for the complexities of interpersonal interactions that may arise in their work.[215, 267] Effective use of adult education methods in training emphasizes both practical skills and theoretic frameworks such as social cognitive theory.

Despite the advantages of state or national standards for CHW workforce development, as of this writing, no national certification for CHWs yet exists.[275] Several states are developing legislation and protocols for CHW training and certification.[277] In the absence of state certification programs, training tends to be designed by the organizations that employ CHWs, primarily to focus training on specific tasks. Hence, CHWs may require additional training if they move to other organizations or disease-associated populations.

An approach to help garner broad support, alignment, standardization, and recognition with widely recognized workforce training would be to collaborate with institutions such as the American Public Health Association, Health Resources and Services Administration within the U.S. DHHS, and major SCD community organizations. A common training curriculum would create a national standardized pool of knowledgeable and skilled CHWs to support the work for SCD care, supplemented by components that address unique populations of or goals for SCD. Beginning in 2015, a CHW-focused Health Resources and Services Administration–funded project through the SCD Association of America spans four regions and at least nine states.[278]

In conclusion, as a rare but socially costly condition that increases health disparities, SCD is in need of alternative interventions for improving short- and long-term outcomes and quality of life. CHWs are an accepted model for improving healthcare outcomes in underserved communities and populations at high risk for poor outcomes. For chronic conditions, CHWs have been shown to bridge cultural, social, and linguistic barriers to chronic disease treatment and improve health outcomes.

Projects underway by members of this group are currently examining the impact of CHW involvement in different dimensions and outcomes of CHW intervention for SCD with diverse targets for specific populations (e.g., adolescents), settings (e.g., community; urban or

rural), and specific healthcare outcomes (e.g., medication adherence). Although results are not yet available, the high rate of patient acceptance of CHWs is an early indicator that CHW interventions can be feasible. Partnership with local and national community-based organizations is emerging as a strong emphasis for SCD, and the SCD Association of America is planning to officially endorse the use of CHWs for SCD.

A CHW SCD intervention research agenda is needed to move the field forward. The authors recommend that efforts to optimize CHWs in SCD be aligned in terms of training, intervention design, and outcomes measurement as well as culturally appropriate alignment with patient medical, social, and mental health needs. The authors propose a national effort to develop best practices for CHWs in SCD and to demonstrate outcomes on the key indicators described above for medical, social, mental health, and family well-being.

A new consensus on roles, training, and certification for CHWs is currently under development by the National Area Health Education Center Organization, funded by the Amgen Foundation, with support from the American Public Health Association. CHWs will be ranked in tiers based upon level of formal education (high school, college), disease-specific education, experience serving as CHW, and experience as supervisor or trainer of other CHWs. These tiers will also correspond to levels of certification and compensation.

3.9. **Implication for Social Welfare Counselling/ Solution and Recommendations**

Counselling and prevention of causes and infections are simple measures not readily accessible to most patients. As a result, the majority of children with the most severe form of the disease die before the age of five, usually from an infection or severe anaemia.

The survivors remain vulnerable to exacerbations of the disease and the complications mentioned above. SCD has major social and economic implications for the affected child as

well as the family. Recurrent sickle-cell crises interfere with the patient_s life, especially with regard to education, work and psychosocial development.

The prevention and control is indeed what can eradicate and combat the disease in this century. Unfortunately, the area of sickle cell disease has been known many decades ago, but people don_t seem to show interest in the eradication. It has drained the purses of many and destroyed many homes and yet people show nonchalant attitude about it. Hundreds and thousands are dying daily due to this incurable disease. It is right time to educate and create awareness for people to know about sickle cell disease and avoid it like a plague, just as people avoid HIV/AIDS today. All newly born babies should be tested for sickle cell disease, because all forms of sickle cell disease are inherited. The test should be done in recognized and well-equipped medical laboratories or any health institutions. Children inherit the genes for the disease from their parent, hence there is need to encourage everyone to know more about it. Eradication of sickle cell disease now depends on the awareness of the disease and guide against the elongation by the masses. The awareness should be done in various organizations such as religious, social, health and academic sectors of our society. Specifically, SCD can be eradicated through the following measures.

Public Education

Accurate, current and clearly written information about sickle cell disease should be produced and made widely available in a credible form, to people. Different forms of communication can be used such as mass media, newspapers, magazines, flier, seminars and workshops in schools, hospitals, organizations, among others. Parents should be educated to educate their wards to know the danger inherent in having children that are sickle cell disease carriers. They should advice their wards not to allow love to blindfold them into future problems, whereby they will not be able to enjoy their marriage as a result of offsprings that will make go in and out of the hospital. Parents can cite examples and educate their children

with vivid experiences of people involved in this type of problems. The public awareness should start from the grass root, involving the NGOs and local government. There are three things that are extremely hard, steel, diamond and knowing one self. Public self-consciousness has good effect on people. As with situationally induced public self-awareness, persons who are high in public self-consciousness tend to be more concerned about how others judge them [279] and are more likely to withdraw from embarrassing situations than those low in this trait [280]. The tendency to comply with external standards encompasses physical appearance as well. A number of studies have found out that individuals high in public self-consciousness are more concerned about their physical appearance and believe appearance is important for smooth societal interaction [281].

There should be outpatient clinic which should provide a setting for patients to be seen by a physician specializing in sickle cell disease. Clinic care should include the services of nurses and certified counsellors as well as a social service assessment. Recommendations for care to be coordinated with the patient primary care physician. And follow up visit should be scheduled for medical management and pain control. Partnerships should be fostered between health professionals, patients, parents, relevant community interest groups and the media, where appropriate. Partnerships will facilitate public education, identification of genetic risks in the community by recording family disease histories, genetic counselling, awareness and active participation prevention and care programmes.

Nevertheless, the dissemination of information can be made in a very simple language that people will understand the concept of sickle cell disease. They can be informed in this manner:

What is sickle cell disease? It is a genetic disorder that results in abnormally shaped red cells. It is inherited from

> the genes of the parents that are carriers. These distorted cells live only 10 – 20 days, compared to normal red blood cells which live about 120 days. This chronic undersupply of red blood cells makes the sickle cell patient anaemic. When the cells clump together, they cause blockages leading to a very severe pain, tissue and organ damage.

The above assertion can be meaningful to the individuals. They should also be intimated with its economic waste, the life of the individual involved is under jeopardy coupled with daily stress.

Transition Programme

To expel this problem from our society, the system of catching them young can be adopted. This is better done at the beginning of late childhood, which is usually the beginning of secondary school education in many societies. To make the awareness rooted, it can be inculcated in the curriculum of the secondary school students especially in their biology subject, but the area of sickle cell disease should be emphasized. They should also be made to understand the implication of the disease in daily living. The awareness should be thorough at the tertiary level because it is actually from this stage many of the youngsters meet their spouse. Proper counselling is very necessary at this stage.

Premarital Counselling

Marriage institution is the bedrock of every society. Premarital counselling is the type of counselling given to youths or couple-to-be to guide against what can erupt in marital life which can lead to endured marriage and even divorce as the case may be. Among many

things that can lead to marriage breakdown is health issue. If the marriage is producing unhealthy children, peace and joy that a marriage is meant to witness will be evaded [503].

Premarital counselling is better given early enough before the two love birds get so enlarged, becoming inseparable as they usually perceive it. Parents should also be bold enough to take their wards for premarital counselling before it is too late. The Counsellor will discuss so many issues in which blood compatibility in terms of genotype can be expatiated. This is a serious issue that the parents should not handle it with levity. It is better for the parents of the two to take their wards for the test themselves in order to avoid deceit. There are also some clergy that are trained Counsellors who should be ready to use their expertise to save mankind. The truth would have to be said whether sweet or bitter if the couple-to-be have AS as their genotype, they should be made to understand the repercussion, the high probability of having children with sickle cell disease and the associated problems, with the pains they are likely to encounter in the area of rearing children. Many of our youths are love intoxicated. The advice may not be meaningful to them, but the experiences of other people that have suffered such menace can be shared with them.

Speakers' Bureau

Sickle cell disease patients can come together forming a body. The organization can choose a specific month for their annual meeting, just like the Breast Cancer Awareness (BCA), Prostate Cancer, Cervical Cancer, HIV/AIDS, to mention a few among others. There should be sickle cell programme in some of our medical centres to provide a variety of services to assist patients and their families in understanding and living with this genetic disorder. In this forum, specialized physicians can educate them on coping strategies and more importantly equipped with firsthand information to enlighten the masses on prevention of the disease. This forum should not encourage self-pity or sympathy but frank talk that would help the

victims to cope and also discourage others falling victims. This kind of seminar should be given wide publicity and making it lively but ensuring that it passes across the information it is expected to pass to the masses. Clubs, individuals, religious organization, both educational and health sectors can be called upon for moral and financial support.

4. Summary and Conclusions

Sickle cell disease is a major public health disease worldwide. There is still a high burden of the disease in Nigeria. There is still a significantly high rate of SCD complications and mortality among Nigerian patients. Current evidence suggests that available care is suboptimal. Largely speaking, prevention, control, and treatment of SCD in Nigeria are still in infancy. Yester efforts albeit present measures appear meager in the face of the enormous disease burden. There is need for a better coordinated effort towards control of SCD by the government at all levels and other concerned stakeholders.

Appropriate interventional programmes backed by an effective national policy should be instituted. In addition, physicians involved in the care of SCD patients should be conversant with current knowledge and standard practices in the treatment of sickle cell disease in order to improve treatment outcomes.

Sickle cell disease is not infectious or contagious, but with all seriousness, it can be eliminated. Taking the above steps into serious consideration, Nigeria can be in the forefront among black nations to fight and eradicate the deadly disease. The sickle-cell disease which has been on rampage for many centuries in Africa can be curbed in Nigeria within the next decade in this millennium.

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This is to certify that the discouse by **Omolara Oluwafunmito ADENIYI** with matriculation number: **LCU/PG/002669** in the Department of Social Work, Faculty of Management and Social Sciences, Lead City University, Ibadan is in full compliance with the approved University Format and Style.

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