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The 63rd session of the UNITED NATIONS (UN) General Assembly held in December 2008 described Sickle Cell Disease (SCD) as “the world’s foremost and lethal genetic disease” which requires global efforts “to bring the disease out of the shadows” and thus adopted a resolution on the, “recognition of Sickle Cell Disease (SCD) as a public health problem” and therefore urged member states to raise awareness as well as set aside 19th June every year as a special day dedicated to all those affected.⁽¹⁾ These include the patients/clients, their families, relatives, health care providers, administrators, government representatives and the international communities.

Sickle Cell Disease (SCD) initiatives in the United States began in the 1960s with the subsequent passage of the National Sickle Cell Control act in 1972 and support provided to set up SCD clinics to conduct; Screening and counseling programmes as part of existing public health care operations; Education and information dissemination activities to both the health care personnel and the general public; Research in the diagnosis, treatment, and control of sickle cell anaemia ⁽²⁾. Work on genetic epidemiological data was initiated for World

Health Organization (WHO) and further developed in United Kingdom by the Genetic Education Programme of the United Kingdom National Screening Committee.⁽³⁾ In summary, most programmes in the developed countries have been successful and have demonstrated that where they exist, survival of patients is steadily improving, affected births are reducing and an increasing number of patients are stabilizing.⁽⁴⁾

The management of SCD in most African countries remains inadequate and SCD control programmes do not exist ⁽⁴⁾. The public health impact/implications of SCD are significant ⁽⁴⁾ It is associated with a high morbidity and mortality as well as a significant socio-economic burden. The need for National SCD control programmes in every country is long overdue ⁽⁴⁾. The role of the programme should be to provide a comprehensive approach to the prevention and management of SCD and thus should be able to utilize simple, affordable and accessible technology that is feasible so as to benefit a large proportion of the community.^(2,3,4) The health care systems should be able to provide basic requirements while education and research activities

provide evidence based practice to fill the knowledge gaps.^(2,5)

Creation or strengthening of SCD control programmes within frameworks of national programmes for prevention and control of non-communicable diseases is necessary in affected countries. This can be made possible through concerted efforts by the government and the Ministry of Health with support from development partners, teaching and research institutions through collaborations^(2,5)

The academic mission of most medical training institutions revolves around EDUCATION, CARE AND RESEARCH.⁽⁵⁾ The mission on EDUCATION in the SCD programme should first entail training of health care workers on prevention, diagnosis and management, then public education on awareness of genetic risks and carrier detection before marriage or pregnancy. The mission on patient CARE should first entail establishment of regional working group experts to coordinate activities, develop guidelines and supervise the activities as they work closely with primary care providers. Ensuring that the health care systems provide the basics for the patients is crucial as well as setting up neonatal and carrier detection screening activities. The mission on RESEARCH should ensure that vital statistics reporting systems guide changes in health policy as well as planning and evaluating appropriate interventions^(2,4,5).

The first description of sickle cells was in 1910 by a Chicago cardiologist and professor of medicine, James B Herrick (1861 – 1954) whose intern Ernest Edward Irons (1877 – 1959) found ‘peculiar elongated and sickle-shaped’ cells in the blood of a dental student at the Chicago College of Dental Surgery who was admitted to the Chicago Presbyterian hospital in 1904 suffering from anaemia.⁽⁶⁾ The disease was named ‘Sickle

Cell Anaemia’ by Verne Mason in 1922 who was then a medical resident in John Hopkins hospital.⁽⁷⁾ Since then a lot of discoveries have been made with regard to the cell biology, genetics, epidemiology, clinical features and management of the sickle cell syndrome.

Sickle Cell Disease (SCD) or Sickle Cell Anaemia (SCA) is a genetic or hereditary blood disorder characterized by red blood cells that assume an abnormal rigid sickle shape (sickling) in conditions of low oxygen concentration like dehydration, acidosis, infection, etc.^(7,8) Haemoglobin comprises four globin chains: - There is the Fetal haemoglobin (HbF) which has two alpha and two gamma chains ($\alpha_2 \gamma_2$) while the Adult haemoglobin (HbA) which has two alpha and two beta chains ($\alpha_2 \beta_2$). Genes in the α -globin and β -globin gene clusters on chromosome 16 and 11 control globin chain production. Due to point mutation, haemoglobin gene variants are present and fall into two broad groups – structured variants in which there is a change in the amino acid sequencing producing unusual haemoglobin (haemoglobinopathy) and thalasaemia in which there is a lower or abolished production of globin chains.^(8,9) Sickle cell anaemia is caused by a point mutation in the β -globin chain of haemoglobin causing the hydrophilic amino acid glutamine to be replaced by the hydrophobic amino acid valine at the sixth position.⁽⁷⁾

The allele responsible for sickle cell anaemia is autosomal recessive and is found on the short arm of chromosome 11.^(7,8,9) Any person carrying the sickle cell gene can transmit it to his or her offspring. Two categories of such persons exist. One category is those who have inherited the gene from one parent and are therefore referred to as carriers or have the sickle cell trait (SCT). They lead normal lives, do not

show any signs of the disease and are often unaware that they carry the gene. The other category is those who have inherited the gene from both parents and are referred to as having sickle cell anaemia (SCA). They have lifelong symptoms and signs of sickness associated with complications of the SCD syndrome.^(7,8,9,10)

A person who has the normal haemoglobin is referred to as having the Hb AA, while one who is a carrier is referred to as having Hb AS and the one who has the disease is referred to as having Hb SS.^(7,8,9,10)

The likelihood that a pregnancy will result into an offspring with sickle cell anaemia is 25 percent if both parents are carriers (Hb AS & Hb AS), 50 percent if one parent is a carrier and the other is a sickler (Hb AS & Hb SS) and 100 percent if both parents are sicklers (Hb SS & Hb SS). It is therefore advisable that those who carry the sickle cell gene (Hb AS or Hb SS) get spouses who have normal haemoglobin (Hb AA) so that their offspring can have either the normal haemoglobin (Hb AA) or the trait (Hb AS)^(10,11)

		Percentage Chance Per Pregnancy (offspring)		
Hb Variant		Normal Hb AA	Carrier Hb AS	Disease Hb SS
Hb AA	Hb AA	100	-	-
Hb AA	Hb AS	50	50	-
Hb AA	Hb SS	-	100	-
Hb AS	Hb AS	25	50	25
Hb AS	Hb SS	-	50	50
Hb SS	Hb SS			100

SCD affects millions of people throughout the world. It is particularly common among people whose ancestors lived in tropical and sub-tropical Sub-Saharan regions. The highest prevalence is among people of African, African-American, Mediterranean (Italian, Sicilian, and Greek), Middle Eastern, East Indian and Central or South American descent.^(12,13) Three quarters of SCD cases occur in Africa.⁽¹⁴⁾ It affects up to about 3 percent births and it is estimated that 6 to 9 million infants are born with SCD in Africa each year.⁽¹⁴⁾ The carrier frequency ranges between 10 to 40 percent across equatorial Africa decreasing to 1 to 2 percent on the North African coast and less than one percent in South Africa.^(14,15) Majority of persons especially in areas with a high burden have the trait (SCT or HbAS), do not know their sickle cell status and get shocked

when they get offspring with sickle cell anaemia.⁽¹⁵⁾ It is therefore advisable that efforts are put in place for people in high burden areas to have their 'sickle cell status' determined.^(2,3,4)

Tests available to determine the 'sickle cell status' include the sickling test, Hb Electrophoresis, Capillary Electrophoresis, Isoelectric Focusing, High performance Liquid chromatography, Tandem mass spectrometry, Immunoassay (sickle SCAN) among others.^(4,16,17) Most of these tests are expensive and not easily available or accessible to majority of Kenyans. The sickle SCAN test which is designed to give results within five minutes at the point of use is promising.^(4,16,17)

The intervention strategies for SCD are categorized into primary, secondary and tertiary by the World Health Organization

(WHO).⁽¹⁸⁾ The tertiary intervention entails managing the complications (anaemia, vaso-occlusion and chronic organ damage) as the patients present with the signs and symptoms.⁽¹⁸⁾ The secondary intervention entails screening the newborns, identifying those with SCD, follow them up and provide prophylactic management in terms of giving standard vaccines as well as other special ones (Pneumococcal and Typhoid), haematinic supplementation (folate) and prophylaxis against malaria and other infections.⁽¹⁸⁾ The primary intervention entails preventing birth of sicklers by ensuring persons are aware of their carrier status, there is pre-conception genetic counseling and reproductive choices are made.^(4,18,19)

In order to be effective, all these intervention strategies should be carried out concurrently in an organized manner and in a SCD control Programme.^(4,18,19) The first essential step towards addressing the burden is to create a SCD registry or data base both at County level and National level, identify the patients either clinically or through newborn screening and profile them in a standard format so as to help plan for their comprehensive care.^(4,18,19) Another step is to set up services which can provide

the appropriate facilities: for determination of haemoglobin variants so that individuals can know their 'sickle cell status'; Public health education on prevention especially pre-conception genetic counseling and reproductive choices.^(4,18,19) The other very important step is to have an organized way in which all the relevant stakeholders can provide their support in terms of guidelines and policies, technical and expert services, funds for drugs, equipment and personnel.^(4,18,19)

The role of a universal National Health Insurance Fund (NHIF) in the provision of an essential health care package in terms of curative and preventive elements to patients with SCD and their families is a special support which all stakeholders should embrace.⁽²⁰⁾ Research activities done should provide sound evidence to convince policy makers of the feasibility and benefits of offering services through the programme.⁽⁵⁾

The WHO recommendation that at least 50 percent or member states should have established SCD control programmes by 2020 is a dream that should be realized by all African countries including Kenya and indeed TEAM (Together Each one Achieves More) work by all stakeholders is necessary in this situation.^(1,2,4,18)

SICKLE CELL DISEASE REGISTRY FORM	
Date : / /	
Health Facility:	Is the patient a referral? <input type="checkbox"/> No <input type="checkbox"/> Yes if yes, indicate referring facility _____
1. Demographic Information	
First name:	Middle name Last name:
Serial Number:	Telephone Contact:
Hospital Number:	Other Number:
D.O.B: / / Estimated Age Years	Gender <input type="checkbox"/> Male <input type="checkbox"/> Female
County:	Sub County/Division/Constituency:
Location:	Sub-location:

Village:		Residence:	
Family name/clan name:		Landmark: nearest Sch/Hosp/Church	
2. Subject occupation <input type="checkbox"/> N/A(Child) <input type="checkbox"/> Student <input type="checkbox"/> Unemployed <input type="checkbox"/> Informal employment <input type="checkbox"/> Formal /Professional employment Specify			
3. Person accompanying patient: <input type="checkbox"/> Mother <input type="checkbox"/> Father <input type="checkbox"/> Sibling <input type="checkbox"/> Children's Home <input type="checkbox"/> Grandparent (<input type="checkbox"/> Paternal <input type="checkbox"/> Maternal) <input type="checkbox"/> Auntie (<input type="checkbox"/> Paternal <input type="checkbox"/> Maternal) <input type="checkbox"/> Uncle (<input type="checkbox"/> Paternal <input type="checkbox"/> Maternal) <input type="checkbox"/> Spouse <input type="checkbox"/> Self <input type="checkbox"/> Other:			
4. Maternal information			
Name (Initials) :		Date of Birth (DOB) : / /	<input type="checkbox"/> Alive <input type="checkbox"/> Dead
Highest level of education: <input type="checkbox"/> None <input type="checkbox"/> Primary completed <input type="checkbox"/> Secondary completed <input type="checkbox"/> Tertiary completed			
Employment status: <input type="checkbox"/> Housewife/Household <input type="checkbox"/> Self-employment (Specify) _____ <input type="checkbox"/> Formal /Professional employment(specify): _____			
<input type="checkbox"/> Other (specify all):			
Marital Status: <input type="checkbox"/> Married monogamous <input type="checkbox"/> Married polygamous <input type="checkbox"/> Single <input type="checkbox"/> Separated <input type="checkbox"/> Widowed			
5. Paternal information			
Name (Initials) :		Date of Birth (DOB) : / /	<input type="checkbox"/> Alive <input type="checkbox"/> Dead
Highest level of education: <input type="checkbox"/> None <input type="checkbox"/> Primary completed <input type="checkbox"/> Secondary completed <input type="checkbox"/> Tertiary completed			
Employment status: <input type="checkbox"/> None/Peasant farmer <input type="checkbox"/> Self-employment (Specify) _____			
<input type="checkbox"/> Forma/ Professional employment(specify): _____			
<input type="checkbox"/> Other (Specify all):			
Marital Status: <input type="checkbox"/> Married monogamous <input type="checkbox"/> Married polygamous <input type="checkbox"/> Single <input type="checkbox"/> Separated <input type="checkbox"/> Widower			
6. Family Information			
Number of siblings:		Birth order(patient) :	sibling survival status: <input type="checkbox"/> No. Alive <input type="checkbox"/> No. Dead
Are you under any medical cover: <input type="checkbox"/> No <input type="checkbox"/> Yes <i>If Yes,</i> <input type="checkbox"/> NHIF <input type="checkbox"/> Other(specify):			
Is it currently active and paid for: <input type="checkbox"/> No <input type="checkbox"/> Yes Does it cover sickle cell as an essential package? <input type="checkbox"/> No <input type="checkbox"/> Yes			
Date of enrollment (medical cover) : / /			
Mode of enrollment: <input type="checkbox"/> Automatic/Place of work <input type="checkbox"/> Direct remission to fund <input type="checkbox"/> Other(specify):			
Any relative(s) ever diagnosed with sickle cell disease? <input type="checkbox"/> No <input type="checkbox"/> Yes <i>If Yes, how many?</i> _____			
Relationship	Name (initials)	Alive	Dead
1.		<input type="checkbox"/>	<input type="checkbox"/>
2.		<input type="checkbox"/>	<input type="checkbox"/>
3.		<input type="checkbox"/>	<input type="checkbox"/>
4.		<input type="checkbox"/>	<input type="checkbox"/>
5.		<input type="checkbox"/>	<input type="checkbox"/>
7. Patient' Medical Information:			
Date of diagnosis of sickle cell disease: / /		Age at first diagnosis:	
Mode of diagnosis of sickle cell disease (tick all that apply):			
Comments			
<input type="checkbox"/> Clinically _____			
<input type="checkbox"/> Sickling test _____			
<input type="checkbox"/> Hb Electrophoresis _____			
<input type="checkbox"/> Other Specify: _____			

Ever had hospital admissions?		<input type="checkbox"/> No	<input type="checkbox"/> Yes If yes?
	Number	Comments	
Last 12 months	_____	_____	
>12< 24 months	_____	_____	
> 24 months ago	_____	_____	
Ever received blood transfusion?		<input type="checkbox"/> No	<input type="checkbox"/> Yes if yes, explain
	Number	Comments	
Last 12 months	_____	_____	
>12< 24 months	_____	_____	
> 24 months ago	_____	_____	
Ever received any of the following vaccines?			
		<input type="checkbox"/> No	<input type="checkbox"/> Yes
Pneumococcal		<input type="checkbox"/> No	<input type="checkbox"/> Yes
Typhoid vaccine		<input type="checkbox"/> No	<input type="checkbox"/> Yes
Yellow fever vaccine		<input type="checkbox"/> No	<input type="checkbox"/> Yes
Meningococcal vaccine		<input type="checkbox"/> No	<input type="checkbox"/> Yes
Other specify		<input type="checkbox"/> No	<input type="checkbox"/> Yes
Ever used the following medications?			
	Before	currently	Date started
			Date stopped
Folate	<input type="checkbox"/>	<input type="checkbox"/>	/ /
Pen V	<input type="checkbox"/>	<input type="checkbox"/>	/ /
Hydroxyurea	<input type="checkbox"/>	<input type="checkbox"/>	/ /
Other specify:	<input type="checkbox"/>	<input type="checkbox"/>	/ /

Where do you attend clinic? _____

Where would you like to attend clinic? _____

Any other relevant information?

FORM FILLED BY (initials& sign) _____ DATE _____

COMPUTER ENTRY DONE BY _____ DATE _____

HAEMOGLOBIN VARIANT SREENING FORM					
Date : / /					
Screening Site:		Is the client a referral? <input type="checkbox"/> No <input type="checkbox"/> Yes if yes, indicate referring facility _____			
1. Demographic Information					
Serial Number:		Telephone Contact:			
Hospital Number:		Other Number:			
D.O.B: / /	Age (years)	Gender		<input type="checkbox"/> Male	<input type="checkbox"/> Female
County:		Sub County/Division/Constituency:			
Location:		Sub-location:			
Village:		Residence:			
Family name/clan name:		Landmark: nearest Sch/Hosp/Church			
2. Subject occupation <input type="checkbox"/> N/A(Child) <input type="checkbox"/> Student <input type="checkbox"/> Unemployed <input type="checkbox"/> Informal employment <input type="checkbox"/> Formal /Professional employment Specify					
5. Family Information					
Number of siblings:			Birth order(client) :		
Are you under any medical cover: <input type="checkbox"/> No <input type="checkbox"/> Yes If Yes, <input type="checkbox"/> NHIF <input type="checkbox"/> Other(specify)?					
Date of enrollment (medical cover) : / /					
Mode of enrollment : <input type="checkbox"/> Automatic/Place of work <input type="checkbox"/> Direct remission to fund <input type="checkbox"/> Other(specify):					
Any relative(s) ever diagnosed with sickle cell disease: <input type="checkbox"/> No <input type="checkbox"/> Yes <i>If Yes, how many?</i> :_____					
Relationship		Name (initials)		Alive	Dead
1.				<input type="checkbox"/>	<input type="checkbox"/>
2.				<input type="checkbox"/>	<input type="checkbox"/>
3.				<input type="checkbox"/>	<input type="checkbox"/>
4.				<input type="checkbox"/>	<input type="checkbox"/>
5.				<input type="checkbox"/>	<input type="checkbox"/>
Client's Screening Test Results:					
IsoElectric Focusing		Hb Electrophoresis		Other (Specify)	
<input type="checkbox"/> Hb AA		<input type="checkbox"/> Hb AA		<input type="checkbox"/> Hb AA	
<input type="checkbox"/> Hb AS		<input type="checkbox"/> Hb AS		<input type="checkbox"/> Hb AS	
<input type="checkbox"/> Hb AC		<input type="checkbox"/> Hb AC		<input type="checkbox"/> Hb AC	
<input type="checkbox"/> Hb SS		<input type="checkbox"/> Hb SS		<input type="checkbox"/> Hb SS	
<input type="checkbox"/> Hb SC		<input type="checkbox"/> Hb SC		<input type="checkbox"/> Hb SC	
<input type="checkbox"/> Hb CC		<input type="checkbox"/> Hb CC		<input type="checkbox"/> Hb CC	
<input type="checkbox"/> Other (Specify)		<input type="checkbox"/> Other (specify)		<input type="checkbox"/> Other(specify)	

Any other relevant information?

FORM FILLED BY (initials& sign) _____

DATE _____

COMPUTER ENTRY DONE BY _____

DATE _____

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