DRAFT STRATEGIC PLAN

2015-2020

THE SHIKURI PROJECT

SAVING FROM THE SCOURGE OF SICKLE CELL DISEASE

SILVER SPRING, MARYLAND

NYANZA/WESTERN /Coastal Regions, KENYA

I. Executive Summary

The Shikuri Project is a registered Non-Profit corporation (501c3) in the United States, dedicated to providing sustainable, Chronic Health Care Management (CHCM) programmes to reduce mortality and morbidity of young children with Sickle Cell Disease, in underserved communities in Kenya, and eventually East Africa. It will achieve the expected results primarily through technical-assistance project management with heavy reliance on partnerships including South-South technical cooperation. Sickle-cell disease (SCD), an inherited disorder of the red blood cells, is the most prevalent genetic disease in Sub-Saharan Africa (SSA). The World Health Organization (WHO) estimates that 50%–80% of infants born yearly with SCD in Africa die before the age of five years, which is the most vulnerable period of the disease. While death is usually from an infection, stroke, or severe anaemia, the complications of the disease affect and kill young children before doctors even suspect that they have the disease.[1] Survivors suffer end-organ damage which shortens their lifespan, and they remain vulnerable to exacerbations of the disease and the complications mentioned above. Persons with SCD are often stigmatized, and SCD has major socioeconomic implications for affected persons, families, communities and nations. The good news is that WHO estimates that 70% of SCA deaths in Africa are preventable with simple, cost-effective interventions such as early identification of SCA patients by newborn screening (NBS) and the subsequent provision of comprehensive care. Screening programs for SCD, initiated in the United States and Jamaica, have led to sharply reduced mortality and morbidity of the disease in young children. Despite the fact that the United Nations has declared SCD as a Public Health Priority for SSA, as an important cause of child mortality in many African countries, there is negligible investment in public health programs. In SSA adequately trained health professionals, specialized health care facilities, effective medicines, vaccines and safe blood transfusion are very limited. There is a dearth of reliable data which can perhaps account for the lack of attention to the disease. In Kenya there are no well- structured care services for early SCD diagnosis and follow up in the highest affected areas. Since 60 % of birth deliveries in the endemic areas do not occur in health facilities, the target group has negligible access to, and is beyond the reach of health care management services. .

The Shikuri Project is therefore establishing a pilot CHCM project in 2 underserved communities of Kenya where the disease has highest prevalence. It will deliver 4 Deliverables of the WHO Strategy for Sickle Cell Disease designed to reduce infant mortality and suffering by at least 50 percent. The Shikuri Project will function synergistically with national and international partner organizations within Kenya's Quality Model for Health. Based on the success of the pilot project, the programme will expand to other communities. A fund-raising and marketing strategy will be developed for project operations, and to advocate for its goals. This will target access to grants and expertise from Foundations, other Not for Profit organizations, Government agencies, and donations from the general public. Please join us to raise resources and implement this programme to save the lives and promote greater wellbeing of affected children, their caregivers and communities.

II. *Mission Statement*

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[1] Newborn Screening for Sickle Cell Disease in Ghana, Kwaku Ohene-Frempong, M.D.,<General News of Ghana, March 2005

1. *The Shikuri Project* exists to introduce and implement sustainable, Chronic Health Care Management (CHCM) programmes to reduce the number of deaths and painful crises suffered by young children with Sickle Cell Disease, in underserved communities in Kenya, and eventually East Africa.

III. Vision Statement

2. The Shikuri Project envisions the establishment of sustained, Sickle-Cell diagnostic and treatment Centres and trained personnel serving communities with high -prevalence and lack of access to national programmes in East Africa.

IV. ***The Entity***

3. *The Shikuri Project* is a Non-Profit corporation with 501 c3 status in the U.S.A., established in February 2014 and awaiting accreditation in Kenya. It is, accordingly, bound by and functions within the respective legal and institutional frameworks and by-laws. Headquartered in Maryland, USA its institutional capacity will be at full strength by the first quarter of 2015. Operations will be based in Kenya for Phase I with expansion in later phases to other parts of East Africa. Initial capital totals US$20,000. *Given the voluntary nature of the involvement of the principals and executive, the Shikuri Project* will function with minimal administrative overhead. In-kind and financial resources are anticipated to be mobilized through its President, Volunteers, and Advisory Board from Donors, Foundations, Grants and the general public. The first Pilot project will initiate activities by January 2015.

4. The Management Structure comprises the President, Secretary, Treasurer and Advisory Board. Gail Sealy, the Founder and President, is a retired United Nations Official and the Mother of Shikuri Sealy, after whom *The Shikuri Project* is named (*About Shikuri*[2]*)* Gail has had a 20-year career with the United Nations in International Development, including serving as Sr. Advisor for Institutional Development with the World Health Organization (WHO)/ Pan American Health Organization from 2006-2008. She therefore brings rich expertise, commitment, and networks in the planning and delivery of human-development services. Her experience of the challenges, blessings, and rewards of parenting and caregiving a child afflicted with SCD has given Gail a deeper, empirical perspective and commitment. In light of the lack of attention to this disease, Gail and her son wish to provide broader humanitarian service to other similarly afflicted children who are beyond the reach of conventional medical care through the Project. The (unremunerated) Secretary is Lilian Thairu, a Kenyan National, who is Organization Development Analyst at the UN Development Programme, New York. The interim Treasurer is Mr. Leon Thomas, Financial Consultant and former Treasury Adviser in the United Nations Development Programme, New York.

5. The Advisory Board comprises the following distinguished Professionals

1. Professor Graham Roger SERJEANT, MD

Oct. 1, 99 – present Professor Emeritus, Faculty of Medical Sciences, University of the West Indies, Kingston, Jamaica; Chairman, Sickle Cell Trust, Jamaica.

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[2] Shikuri was born with Sickle Cell Disease (SCD) in Kenya and had not been diagnosed prior to his adoption at the age of 2 years old. Due to her constrained circumstances, his birth mother, who was from Western Kenya, abandoned him in a hospital in the capital city, Nairobi, where he was fortunate to receive blood transfusions. He was later transferred to an orphanage which, unaware of his diagnosis, did not provide him with any medications. He therefore suffered recurrent, untreated pain crises, temporary blindness, and a minor stroke. Since adopting him, Gail, the Founder, was fortunate enough to avail him of state- of- the -art medical care. His hospitalizations for painful crises, life threatening fevers and pneumonias have reduced from 7 episodes per year to one or two; and he is in generally good health... Through "The Shikuri Project", we aim to save and improve the lives of other underserved children in E Africa born with Sickle Cell disease through early diagnosis and sustained treatment.

Honorary Appointments

- 1986-present Chairman, Sickle Cell Trust (Jamaica).
- 1995-1999 Director, WHO Collaborating Centre in Haemoglobinopathies
- 1995-present Advisor to the InterMinisterial Committee of the Brazilian Government on the development of a National Sickle Cell Program
- 1997-2000 President of COSCA (Caribbean Organisation of Sickle Cell Associations).
- 1998 Honorary Fellow, Royal College of Physicians, Edinburgh
- 1999-present Visiting Professor, Department of Infectious and Tropical Medicine, London School of Hygiene and Tropical Medicine, London
- 1999-present Honorary Professor, Department of Public Health, Guy's Hospital, London
- 1999-present Professor Emeritus, University of the West Indies, Kingston
- 2000-present Honorary Consultant, King's College Hospital, London
- 2000-present Honorary Consultant, City Hospital, Birmingham
- 2001-present Honorary Consultant, Homerton Hospital, London
- 2002-present Visiting Professor, Dept. of Paediatrics & Child Health, Mulago Teaching Hospital, Makerere University, Kampala, Uganda
- 2003 Aflac Visiting Professor, Children's Health Care of Atlanta, USA
- 2004 Visiting Professor, Dept. of O&G, Meharry Medical College, Nashville, TN, USA

Over the last 47 years in Jamaica, developed a major comprehensive clinical and research facility for the management and investigation of over 5,000 patients with sickle cell disease, an inherited blood abnormality affecting approximately 400,000 births annually worldwide. By documenting the natural history of this condition and evolving simple methods for the prevention or more effective treatment of complications, the Jamaican Unit has significantly decreased mortality and reduced morbidity of this disease. The methods pioneered in the Jamaican Unit were generally simple, cost-effective, and low technology and hence were appropriate to many countries with limited resources, where the disease is a common public health problem. The Cohort Study, based on the detection and follow-up of 550 cases of sickle cell disease among 100,000 consecutive normal deliveries at Victoria Jubilee Hospital is a unique study of the natural history of sickle cell disease made possible by an island community such as Jamaica. For the first time, the true natural history of the disease could be documented in place of the symptomatic bias inevitable in previous reports. As the children have aged, the major determinants of morbidity and mortality at each age have been defined, and cost-effective interventions implemented. The Jamaican Cohort Study is recognized worldwide as a major contribution to the understanding and improved management of sickle cell disease.

2. Professor Dr William M Macharia

The Hassan Ali Dhanari Professor and Chair
Department of Paediatrics and Child Health, Agha Khan University Hospital, Nairobi, Kenya

3. Dr. Vincent Akoko Orinda, M.D

A senior health consultant, with over 17 yeard' experience with UNICEF and has provided technical support to many countries especially in Africa. He served as a Senior Health Advisor (Child Health) for UNICEF in New York and also worked in Namibia, South Africa and Uganda. Dr. Orinda has a wide experience in health systems (policy and strategy

development) as well as in community based health development. Prior to joining UNICEF in 1990, he was a Senior Lecturer in Paediatrics and Child Health, University of Nairobi.

4. **Professor Babatunde Thomas**
   Managing Director, African Capacity Matters (ACM) Ltd and
   Former United Nations Resident Coordinator, Uganda
   Nairobi
   Kenya

5. Dr Babette Barbash Weksler, M.D.
   - USA Professor Emerita of Medicine, Joan and Sanford I. Weill Department of Medicine, Weill Cornell Medical College, New York, USA 2014 -
   - Professor of Medicine, Joan and Sanford I. Weill Department of Medicine, Weill Cornell Medical College 2007 - 2014
   - Professor of Medicine, Joan and Sanford I. Weill Department of Medicine, Weill Cornell Medical College 1981 - 2007

   Dr. Weksler was the first to observe that endothelial cells were a major source of the protective prostaglandin, prostacyclin

6. **Dr Nicole Kucine**
   Assistant Professor of Pediatrics
   Division of Pediatric Hematology/Oncology Department of Pediatrics
   Weill Cornell Medical College
   New York, NY
   USA

7. **Ms Foulata Kwena**
   Vice Chairperson
   The Kenya Children Sickle Cell Foundation
   Nairobi, Kenya

8. **Dr. Jennifer Knight-Madden, MBBS, Bd Cert Ped Pulm PhD**
   Head, Sickle Cell Unit, University of the West Indies
   Jamaica

9. Mr Hernan Rosenberg
   Public Policy and International Development Consultant
   Former Regional Advisor in Social Protection and Health Investments
   Pan American Health Organization
   Washington, DC

VIII *Resources Generation Strategy*

6. Operating funds and project inputs will be sourced from fundraising and the enrolment of Volunteers. Fundraising will be anchored in and radiate from each Project Deliverable. Given the Board Members' considerable individual expertise, it is proposed that, working in facilitated teams, they will generate Activities, an Implementation Plan, Budget, and (optional) Fundraising options for each Deliverable. Board Members may lead and support teams on more than one Deliverable. The Project is currently recruiting an Advisory Board member whose focus and expertise will be Fund-raising. A fund-raising strategy for Project operations will be developed to market the project, advocate for its

goals, and access grants and expertise from Foundations, other Not for Profit organizations, Government agencies, and donations from the general public.

VIII Situation Analysis And Problem To Be Addressed

7. Sickle-cell disease (SCD), an inherited disorder of the red blood cells, is the most prevalent genetic disease in Sub-Saharan Africa. The World Health Organization (WHO) estimates that approximately, 50%–80% of the estimated 230-400 000 infants born yearly with SCD in Africa die before the age of five years, which is the most vulnerable period of the disease. WHO has therefore declared SCA as a public health priority?

8. Among people with SCD, "sickle" or abnormally shaped red blood cells get stuck in small blood vessels and block the flow of blood and oxygen to organs in the body. These blockages can cause repeated episodes of severe, excruciating pain, organ damage, and serious infections, stroke, and/or death. The process of experiencing pain with this process is known as the sickle cell crisis. The complications of the disease affect and kill young children before doctors even suspect that they have the disease.[3] Presently, there is no failsafe cure for sickle-cell disease. Even in developed countries where stem cell transplantation can be contemplated, there is no widely acceptable public health intervention for the clinical cure of SCD.7

9. SCA is estimated to be currently responsible for more than 6% of all deaths in African children younger than 5 years.4 In many countries, 10%–40% of the population carries the sickle-cell gene resulting in estimated SCD prevalence of at least 2%. Estimates suggest that 6 million Africans will be living with SCA anaemia if average survival reaches half the African norm. Prevalence levels decrease to between 1% and 2% in North Africa and to less than 1% in southern Africa. In countries such as Cameroon, Republic of Congo, Gabon, Ghana and Nigeria, the prevalence is between 20% and 30% while in some parts of Uganda and Kenya (Western and Coastal Regions) it is as high as. 45%. The prognosis of the population with the most severe form of the disease Sickle Cell ("SS") is grim: More than 50% of the children in areas of high prevalence die before the age of five, usually from an infection or severe anaemia;[4] 80% die before adulthood. Exacerbating the risks to the affected population are the dangers of malaria, and pneumonia from the pneumococcal virus. The sickle cell trait has a partial protective effect against malaria. However, ironically, for people with Sickle Cell Disease SS (inherited from the gene of both parents), the effects are the most severe and they are far more likely to die from malaria. Studies on the subject highlight both the central role that malaria plays in the high early mortality seen in African children with SCA and the urgent need for better quantitative data. [5] [6]

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[3] Newborn Screening for Sickle Cell Disease in Ghana, Kwaku Ohene-Frempong, M.D.,<General News of Ghana, March 2005

[4]AFR/RC56/17 SICKLE-CELL DISEASE IN THE AFRICAN REGION: Addis Ababa, Ethiopia, 28 August–1 September 2006

[54] High mortality from *Plasmodium falciparum* malaria in children living with sickle cell anemia on the coast of Kenya hematologylibrary.org/content/116/10/1663.full.html

Charlotte F. McAuley1,2,
Clare Webb1,2,

10. There is a dearth of reliable data which can perhaps account for the lack of attention to the disease. In many Sub-Saharan African countries, the lack of an evidence-base has been a key factor in the limited scale of efficacious interventions, such as penicillin prophylaxis [8]. Most countries in Africa do not have Newborn Screening Programmes (NBS) for SCD; therefore evidence relies on hospital-based studies. Information from such cohorts is biased, as it will on one hand consist of healthy survivors and on the other, will not identify those with mild disease who do not seek healthcare or those with severe disease who have died. This situation is similar to that in Jamaica and USA in the early 1970s, when NBS for SCA was not established and evidence relied on prospective studies in hospital-based cohorts, where most of the patients (92% and 65% respectively) were not identified at birth[4], [9]. Despite the limitations of hospital-based studies, these studies provided important evidence on morbidity and mortality due to SCA.

11. Quantifying the number of under-5 child deaths from SCD in African countries is important to attract policy support and resources for measures to reduce the burden of mortality."[7] In the absence of newborn diagnosis programmes, there is an acknowledged challenge to locally identify children with SCD/SCA. Complicating the difficulties in identifying the target group is the reality that the reporting of infant mortality rates is largely overshadowed by other prevalent conditions,[8] which are often themselves complications of SCD, such as pneumonia, sepsis, and malaria. Ironically, the fatalities caused by SCD are subsumed under the disease's own complications. These are exponentially more documented in research studies and thus the recipient of greater policy and funding attention. Given that resources for health are limited and must be prioritized, it is important to increase awareness to advance it on the public health agenda.

Therapies

12. The good news is that WHO estimates that 70% of SCA deaths in Africa are preventable with simple, cost-effective interventions such as early identification of SCA patients by newborn screening (NBS) and the subsequent provision of comprehensive care. Elevated mortality due to SCD among children younger than 5 years has been virtually eliminated from North America by this means, including the provision of penicillin prophylaxis and polyvalent pneumococcal vaccination. For those patients with

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- Julie Makani[1,3],
- Alexander Macharia[1],
- Sophie Uyoga[1],
- Daniel H. Opi[1],
- Carolyne Ndila[1],
- Antony Ngatia[1],
- John Anthony G. Scott[1,4],
- Kevin Marsh[1,4], and
- Thomas N. Williams[1,2,4,5]

[7] Sickle Cell Disease in Africa A Neglected Cause of Early Childhood Mortality
Scott D. Grosse, PhD, Isaac Odame, MB, ChB, MRCP, Hani K. Atrash, MD, MPH, Djesika D. Amendah, PhD, Frédéric B. Piel, PhD, Thomas N. Williams, PhD

[8] A 2005 US CDC Health and demographic surveillance study in rural western Kenya[8] determined that the infant mortality rate was 125 per 1000 live births, and the under-five mortality rate was 227 per 1,000 live births due primarily to malaria and HIV/AIDS. This is considerably higher than the national average which is 75 per 1,000 live births

access to tertiary facilities, who suffer repeated, severe complications of sickle cell disease, particularly children with a stroke history, simple transfusions or exchange transfusions are often used to preserve organ function and prolong life. [12] Despite the usefulness of chronic transfusion, its long-term effects include iron overload, which can damage the liver. Transfusion therapy is not available to the vast majority of the target beneficiaries who are beyond the reach of tertiary services.
[i]Currently, hydroxyurea is the only disease-modifying therapy approved for sickle cell disease. [15] It has proven to reduce the frequency of painful crises and the need for blood transfusions in patients with recurrent painful crises. Its efficacy is generally attributed to its ability to boost the levels of fetal hemoglobin (Hb F, $\alpha 2\gamma 2$) and to increase the water content of red blood cells, decrease the neutrophil count, and alter the adhesion of red blood cells to the endothelium. *The Shikuri Project*, therefore, places the highest priority on availing the target population of this medication. In malaria endemic areas, given the vulnerability of children living with SCA, studies confirm the importance of providing them with effective prophylaxis. The other important components of treatment are early intervention with analgesics, antibiotics, polyvalent pneumococcal vaccination, folic acid supplementation, and high fluid intake. At times, invasive procedures such as surgery may be needed. There is sufficient evidence that neonatal screening for sickle-cell disease, when linked to timely diagnostic testing, parental education and comprehensive care, markedly reduces morbidity and mortality in infancy and early childhood.

The Challenge of Access to Care

13. In most of the SSA countries where sickle-cell disease is a major public health concern, national programmes for its control do not exist. Basic facilities to manage patients are usually absent, systematic screening for sickle-cell disease is not common practice and the diagnosis of the disease is usually made when a severe complication occurs. Simple, inexpensive and cost-effective procedures such as the use of penicillin to prevent infections are not available to most patients. Where control programmes do exist, these have neither the national coverage nor basic facilities to manage patients. This is the case in Kenya and Western Kenya, in particular. Systematic screening for SCD using a simple blood test is not a common practice, and diagnosis (when made) is usually made when a severe complication occurs and the patient is within reach of health-care facilities. 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 suffer end-organ damage which shortens their lifespan, and they remain vulnerable to exacerbations of the disease and the complications mentioned above. Persons with SCD are often stigmatized, and SCD has major socioeconomic implications for affected persons, families, communities and the nation. Recurrent sickle-cell crises interfere with the patient's life, especially regarding education, work and psychosocial development. Among the target population, knowledge is limited and misconceptions abound on the nature of the disease. Knowledge is negligible about the value of early diagnosis, use of penicillin prophylaxis in childhood, timely treatment of infections and avoidance of fluid depletion. Misconceptions about SCD also leads to stigmatization with health and social consequences.

There are few adequately trained health professionals and specialized SCD management facilities in the country. The situation is further compromised by the erratic availability of important medicines like hydroxyl urea, vaccines like pneumovac and safe blood supply. In some districts of Western Kenya, and the Coast region which have the highest prevalence in Kenya, there are no dedicated treatment centres or health care management systems in place. Amidst the abject poverty that characterizes these populations, affording treatment for many families is a distant dream that manifests in high infant and child mortality. Furthermore,

the majority of the families employed household heads or health insurance cover denying them ready access to health facilities and live on less than a dollar a day.[9] .

14. Despite logistic and economic constraints, neonatal SCD screening along with CHCM have been successfully practised in some parts of Africa, including Benin and Ghana and other developing countries, including Jamaica. For example, in Benin where neonatal screening and CHCM were practised, the under-five mortality rate of SCD was 15.5 per 10 000, which is ten times lower than the overall under-five mortality rate. These findings are consistent with those from developed countries, demonstrating the benefit of newborn screening and close follow-up of children using CHCM.

IX. Strengths and Opportunities: National and Partners 'Response

15. The Republic Of Kenya's Health Policy 2012 – 2030 has the goal to 'attain the highest possible health standards in a manner responsive to the population needs". Sickle Cell Disease (SCD) is not explicitly identified as a priority in The Second Medium Term Plan For Health, the Health Sector Strategic And Investment Plan (KHSSP). However, the following policy focus areas and planned interventions will positively impact the project beneficiaries, who are beyond the reach of current services: (i) the halting and reversing of Non-Communicable Diseases; (ii) Providing essential health care (iii) organization of service delivery with concentration at the community level including the establishment of Monthly Mobile Clinics. The Shikuri Project will function synergistically within the framework of the KHSS), specifically, the Kenya Quality Model for Health. Many of the key health initiatives of International Foundations, WHO, UNICEF, and the Govt of Kenya which have resulted in reductions in infant mortality will have had a positive impact on the survival rates of children with SCA.[10] These include national malarial control programmes, the introduction of both *Haemophilus influenzae* type b vaccine (which has led to a sustained reduction in hemophilus disease15) and of free access to insecticide-treated bed nets for children and pregnant women (to which major declines in malaria incidence have been attributed54) and the planned introduction of the 10-valent conjugate vaccine against *Streptococcus pneumoniae* that covers 80% of the prevailing *S pneumoniae* serotypes.30 ORS sachets for children are available in public health facilities and provided freely to under-fives. In Homa Bay and Siaya Counties, UNICEF is supporting the MOH/County governments to implement Integrated Community Case Management (ICCM) with focus on management of diarrhea, malaria and pneumonia. In some counties, there is real-time monitoring of sick children (under-five) by Community Health Workers (CHWs) using smart phones implemented by KEMRI with the support of UNICEF, Any child under-five with SCD who has malaria, diarrhea or pneumonia will automatically benefit from ICCM. While the challenge to identify and manage children with SCD/SCA remains, the good news is that all sick children in the community whom the CHWs are not able to manage will be referred to health facilities and followed up by CHWs to ensure completion of referral. Hence, ICCM would greatly benefit children with SCA/SCD as their access to care would be significantly increased.

16. Complementing the national government's efforts are those of international development organizations providing development assistance including the United Nations. There are over 530 Civil Society organizations working in the health sector in Kenya. The Shikuri project prioritizes integral and complementary partnerships with these including, primarily, the Kenya Children Sickle Cell Foundation, and other national, and international, CSO stakeholders working on CHCM to meet the needs of the target group. The Project will accordingly prioritize collaboration with official entities particularly at the County level for all phases of implementation. In addition, the Board is invited to propose, review and decide on partnerships with providers of International Development Assistance (IDA) and Civil Society Organizations

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[9] Concept Note: Empowering Kenyans to deal with Sickle Cell Disease (SCD) also known as Drepanocytosis **COUNTRY: KENYA: CHILDREN SICKLE CELL FOUNDATION, 2012**

(CSOs) operating in health in Kenya for implementing Project activities. These include the Global Fund for Aids, Tuberculosis and Malaria; UNICEF, UNDP, WHO, and the Gates Foundation. The project proposes to utilize and enhance the capacity of extant institutions, including those associated with district hospitals, to reach the widest number of inaccessible communities.

X. Target Group and Beneficiaries

17. Given the prevalence of SCD –SS in Eastern Africa and the understanding in global public health circles that the highest rates of childhood mortality, 50%–90%, among African children with SCD are specific to SS.[1–7,10–12] , this project focuses on the target group with Sickle Cell Disease (SS). According to WHO[11], in some parts of Uganda and Kenya **(**Western and Coastal Regions) the prevalence of the Sickle Cell gene is estimated to be as high as 45%. Based on a benchmark national SCD study in Kenya 1990[12], it is understood that the disease is endemic in Western Kenya/Nyanza Regions (and preponderant among the Luo and Luhya ethnic groups) and, to a lesser degree, in the Coast Province (among the Kambes of the Mijikenda group). The target group in Western Kenya/Nyanza is also at the highest risk in the nation for HIV/AIDS and malaria which are preponderant. Cross-sectional Studies conducted by McAuley and Williams in 2009 and 2010 estimated that in Kenya the occurrence of early mortality from SCD (Sickle Cell SS) for children before the age of 13 was 60-90 percent higher than the average child population.

Frequency of Hb SS by age group: selected cross-sectional studies

Barclay (1971)	Zambia (mining	1969–	0–11	2845	1.3	60% excess mortality by
			1–3 years	2200	0.9	
			3–12 years	2306	0.5	
Fleming et	Nigeria (rural	1970–	Newborns	534	2.1	92% excess
			1–4 years	259	0.4	
			5–14 years	637	0.2	
McAuley et (2010),[29] Williams et al. (2009)[41]	Kenya (Kilifi)	1998–	0–11	782	1.0	90% excess age 13 years
			12–23	282	0.35	
			3–5 years	415	0.24	
			6–13 years	3677	0.09	
Danquah et al. (2010)[47]	Ghana (Northern	2002	0–4 years	1266	0.39	70% excess past age 5 years
			5–10 years	842	0.12	
Simpore et	Burkina Faso	1997–	Newborns	HWE	0.25	Data consistent

[11]. SICKLE-CELL DISEASE IN THE AFRICAN REGION: CURRENT SITUATION AND THE WAY FORWARD
World Health Organization, Report of the Regional Director REGIONAL COMMITTEE FOR AFRICA, June 2006

[12] Survey of sickle disease in Kenya.
Authors Aluoch JR, Aluoch LH.
Journal Trop Geogr Med. 1993 Mar;45(1):18-21.

(2002)[40]			Median 9 years	9201	0.13	excess mortality
Desai et (2005)[48]	Kenya (rural	1998–	Newborns	HWE	1.6	Data consistent excess mortality to the surveys
			0–3 years	2774	0.6	
Allen (1992)[49]	The Gambia	1988	Newborns	HWE	1.2	Data consistent excess mortality
			3–8 years	389	0.3	
Cox et al. (2008)[50]	The Gambia	2003	Newborns	HWE	0.8	Data consistent excess mortality
			10–72 months	536	0.3	
Sarr et al. (2006)[51]	Senegal (rural	2002–	Newborns	HWE	0.5	
			2–10 years	432	None	
Saurin (1984)[52]	Senegal (rural Kégoudou)	1970–	Newborns	HWE	1.0	At least 70% mortality, but no information by age
			All ages	596	0.3	

Hb SS, sickle cell anemia; HWE, Hardy-Weinberg equilibrium

High Child mortality in Nyanza and Western Provinces -- the target region of Phase I

18. The 2008/09 Kenya Demographic and Health Survey (KDHS)[13] found that 1 in 7 children in Nyanza Province (149 per 1000 live births) died before attaining his/her fifth birthday as compared to 1 in 20 (51 per 1000 live births) in Central province. This means that a child in Nyanza province was 3 times more likely to die than a child in Central province. Hence, Nyanza province accounted for 33% of all under-five child mortality in Kenya whereas Western province was found to be the second worst performing province with under-five mortality rate of 121 per 1000 live births, accounting for about 24% of under-five mortality. It is evident from the Demographic and Health Survey Reports that Nyanza province has consistently shown the highest child mortality rates in Kenya over the years as is highlighted in the table below.

19. A Government of Kenya report (Nov 2005) reveals that 65% of the population in Nyanza Province (approximately 2.5 million people) lives in abject poverty. More recent studies such as the Kenya National Human Development Report (KNHDP, 2006) confirm that Nyanza Province is among the country's most impoverished regions. In Kisumu town, 66% of the residents live below the poverty line. Suba, Busia, Tana River, etc are listed as some of the poorest districts in Kenya.[14]

20. There are no well- structured, care services for early diagnosis and follow up of Sickle Cell Disease in these areas. Moreover, since 60 % of birth deliveries in these areas do not occur in health facilities, the target group is not formally diagnosed, has negligible access to, and is beyond the reach of health care management services.

21. *The Shikuri Project*, therefore, proposes to locate the First Phase of Operations in Nyanza and Western Provinces, exploiting opportunities for synergistic partnerships with national and CSOs working on health-care delivery systems on the ground. Following the lessons and successes of this pilot, the programme will expand to serve other communities in East Africa. Given that the most vulnerable period of the disease in affected populations is in children under 5-years old, the project will target newborns and children under 5. Subsequent phases of the Project will target the affected population of the Coast District.

XI. *Programme Goals and Deliverables*

A. Goals:

Phase I: (24 months)

[13] Republic of Kenya. Kenya Demographic and Health Survey 2008/09 Report.

[14] **CONCEPT NOTE: "**Empowering Kenyans to deal with Sickle Cell Disease (SCD) also known as Drepanocytosis", **KENYA: CHILDREN SICKLE CELL FOUNDATION, 2012"**

Establish Pilot Programme, integrating and implementing 4 elements of WHO's Strategy for Sickle Cell Disease in Africa and Advocacy deliverable in one unserved community of Western Kenya/the Western and/or and Luo Nyanza and Coast Province regions with estimated target population of 40,000, This will achieve the following results:

i. baseline and surveillance data on child mortality from SCD established from the target populations;

ii. an increase by 100% in the numbers of infants screened/ children diagnosed and treated

iii. Reduction by 60% in infant/child mortality (which may need to be extrapolated due to lack of current baseline data)

iv. Increase by 60 % of parents-caregivers educated;

v. Sustainable community- based system established for chronic health care and pain management.

B. Deliverables

1. Systems established to provide sustainable, routine Early Identification and Screening Of newborns and Infants for SCD.

There is sufficient evidence that neonatal screening for sickle-cell disease, when linked to timely diagnostic testing, parental education and comprehensive care, markedly reduces morbidity and mortality in infancy and early childhood. Ideally, the disease should be identified at birth as part of routine newborn screening or at any subsequent contact the child has with a health facility or trained practitioner. Diagnosing of sickle cell disease (SCD) in newborns and infants allows health workers to educate parents about the special needs of the children and also to begin preventive treatment before the children begin the develop complications of the disease. Early identification can be done by universal screening of all newborns by collecting blood from a heel prick; testing can be done using iso-electric-focusing or high-performance liquid chromatography. Counselling and health education will be provided to parents/care-givers. This deliverable will incorporate Expectant Mothers into the Target group, which will enable early (pre-natal) diagnosis; counselling and seamless follow-up for the provision of post -natal services (including electrophoresis testing). The Project is seeking advice on functioning symbiotically with ongoing initiatives in HIV diagnosis of newborns and prenatal counselling.

2. SSA Surveillance System Initiated.

i. The dearth of reliable data can be attributed to the following factors: (i) the absence of Newborn Screening Programmes (NBS) for SCD, (ii) the reality that 60% of live births in Kenya occur outside hospitals in a research context which relies on hospital-based studies, and (iii) the masking in Policy Research of the true burden of SCD by reports on its complications disconnected from any linkage with SCD itself. . This lack of data exacerbates the neglect of this

disease in policy making and donor-funding forums. This Project output will seek to address this lack of knowledge by providing (i) an estimate of mortality rates to highlight the burden of disease due to Sickle Cell Anaemia (SCA). The data will provide a basis for increasing targeted interventions, highlighting it in policy making agenda, increasing funding, and evaluating their impact. The proposed approach will be to establish a cohort of SCA patients, with a large sample diagnosed at birth and through infancy, and follow them to determine the success of the interventions. (ii) Findings and estimates on typical syndromic presentations (crises) and comorbidities with: Malaria and/or Pneumonia and/or Malnutrition and/or Other critical health variables. This surveillance may be complemented by additional research on the impact of ***ongoing initiatives*** which have resulted in reductions in infant mortality. These include national malarial control programmes, the introduction of both *Haemophilus influenzae* type b vaccine (which has led to a sustained reduction in hemophilus disease15), free access to insecticide-treated bed nets for children and pregnant women (to which major declines in malaria incidence have been attributed54) and the planned introduction of the 10-valent conjugate vaccine against *Streptococcus pneumoniae* . However, as noted in the valuable study, Sickle Cell Disease in Africa A Neglected Cause of Early Childhood Mortality,[15] if it is found that basic public health measures—including improved nutrition and interventions against malaria, pneumonia, and diarrhea—reduce the burden of infectious diseases, it is likely that the absolute burden of mortality attributable to SCD will decrease. However, correlatively, the relative burden as a fraction of all under-5 mortality actually might increase.

3. **Free and/or affordable or free medicines provided to beneficiary group to prevent death, complications, reduce morbidity and suffering.**

These comprise lifesaving and sustaining medicines, such as Hydroxy Urea; prophylactic antibiotics and anti-malarials; folic acid supplementation; essential supplies, such as thermometers and ORT for each household. Given the efficacy of Hydroxy Urea, *The Shikuri Project*, therefore, places the highest priority on availing the target population of this medication.

[15] Sickle Cell Disease in Africa A Neglected Cause of Early Childhood Mortality
Scott D. Grosse, PhD, Isaac Odame, MB, ChB, MRCP, Hani K. Atrash, MD, MPH,
Djesika D. Amendah, PhD, Frédéric B. Piel, PhD, Thomas N. Williams, PhD

4. **In line with the Ouagadougou Declaration[16], Chronic SCD health care management provided and integrated into local and community health- care system .**

This Deliverable focuses on Chronic Health Care Management (CHCM) with interventions adaptable to local needs of communities. Through this objective, the project will build and/or enhance the capacity of community practitioners, facilities for creation of comprehensive treatment centers accessible to the target populations, and implement interventions adaptable to the local needs of communities. The comprehensive treatment centers will provide: comprehensive (including emergency care); provision of medications, including anti-malarials, hydration; administration of specific vaccines; continuous medical follow-up; early detection and management of complications including fevers; patient referral to higher-care centres when necessary; and - of paramount importance - parent/ care giver and patient education and counselling in the foregoing. This output will also create or enhance the capacity of a cadre of community Health practitioners with training in SCD control including prevention, diagnosis and management of cases.

5. **Increased awareness and knowledge of SCD as well as visibility on the public health agenda by community and national stakeholders (e.g., Community Health Committees in target counties)**

The dearth of reliable data is partially addressed by Deliverable 2, on surveillance. In addition, the reporting of infant mortality rates in the target communities is largely overshadowed by other prevalent conditions, which are often themselves complications of SCD, such as pneumonia, sepsis, and malaria. These conditions are diagnosed in greater numbers, exponentially more reported in surveillance and research studies, and thus the recipient of greater policy and funding attention. Given that resources for health are limited and must be prioritized, this Deliverable focuses is important to increase awareness of SCD to advance it on the public health agenda. As noted in the study, "Sickle Cell Disease in Africa, A Neglected cause of early childhood mortality", "Quantifying the number of under-5 child deaths from SCD in African countries is important to attract policy support and resources for measures to reduce the burden of mortality."[17]

XII. *Implementation/ Strategy*

22. The Shikuri Project foresees the project to be implemented within the framework of the Govt of Kenya's ICCM system, working with established Community Health Units

[16]Ouagodougou Declaration, WHO Meeting….to be added

[17] Sickle Cell Disease in Africa A Neglected Cause of Early Childhood Mortality
Scott D. Grosse, PhD, Isaac Odame, MB, ChB, MRCP, Hani K. Atrash, MD, MPH, Djesika D. Amendah, PhD, Frédéric B. Piel, PhD, Thomas N. Williams, PhD

with a workforce of Community Health Workers (CHWs) as well as a committee oriented/trained in basic primary health care needs. The project will provide direct assistance through the provision of in-kind resources, including medical supplies, training and capacity building through South-South and IDA technical assistance

23. Subject to The Advisory Board approval, *The Project* Deliverables will be anchored in and radiate from the creation and enhancement of comprehensive-care centres. These centres will, ideally, build on the capacity of extant clinics, including those associated with district hospitals or ongoing IDA programmes. Those identified (but not yet approached), to date, include: Western Kenya/Nyanza: St Luke's Clinic at united Mall in Kisumu; DFID Clinic in Kakamega County; Alluoch Clinic in Yala; Eldoret, Moi Referral Hospital; Clinic in the county of Homa Bay; Coast Province: Coast General Hospital or Aga Khan Hospital in Mombasa; KEMRI funded Sickle Cell Centre at Kilifi District Hospital at the Kenyan Coast. (Diagram under development).

Pre-Requisites

24. The following elements will need to be in place in order for credible operations to proceed:

i. Updated epidemiological study of Sickle Cell Disease in the target regions

ii. Linkages established with local public health entities in target populations

iii. Situation Analyses of relevant existing private and public health initiatives; capacities of health centres/clinics (paragraph 30); impact of public and IDA funded progammes /operations on target population. . This will definitively ascertain patient populations, institutional capacity strengths/ needs, opportunities for partnership, and better delineation of project needs.

25. A Project team will be established on the ground in consultation with the local authorities.

XIII. *Risks* – To be elaborated

XIV. *Workplan*

SHIKURI PROJECT BI-ANNUAL WORKPLAN 2015-17

Vision: The Shikuri Project envisions the establishment of sustained, Sickle-Cell diagnostic and treatment Centres serving communities with high lack of access to national programmes in East Africa.

Mission: The Shikuri Project exists to introduce and implement sustainable, Chronic Health Care Management (CHCM) programmes to reduce the nu and painful crises suffered by young children with Sickle Cell Disease, in underserved communities in Kenya, and eventually East Africa.

Overall Objective: Establish Pilot Programme, integrating and implementing 4 elements of WHO's Strategy for Sickle Cell Disease in Advocacy deliverable in underserved communities of Western Kenya and Coast Province regions

Activities	Timeframe	Indicators	Responsible	Means of Verification	Resou
Objective 1: To advocate for routine early Identification and screening of newborns and improved care of cases of Sickle Cell Disease Anaemia					
Conduct an assessment of the magnitude of CSD/SCA in Nyanza and Western provinces This study is intended to provide a rational and evidence based information for initial population estimates to inform recommendations on the provision of care continuum defined by level from community to all referral levels. 1.1 **Deliverables** 1. A comprehensive literature review of SCD/SCA in Kenya with focus on Western Kenya 2. Estimation of children with SCD in Western Kenya extrapolating the numbers for Homabay and Kakamega Counties	Nov 2014 – March 2015		CHMT Study Team Shikuri Team	Study report available	Shiku $10,0

3. Recommend appropriate systems to enable early recognition, diagnosis and treatment continuum of sickle cell disease and as to close to communities as possible with reference to findings on health facilities at tiers 1?2-5 in Goal 2 above. 4. Outline current best care practices for SCD in communities, schools and health facilities 5. Generate a data base on: ii. No of centres by comparative capacities and capabilities in diagnosis and treatment of SCD iii. Estimated no of patients with SCD iv. Estimates of typical syndromic presentations (crises) and comorbidities with: a. Malaria and/or b. Pneumonia and/or c. Malnutrition and/or d. Other					
1.1.1 Identify study team	October 2014				
1.1.2 Finalise the study TOR	October 2014				
1.1.3 Conduct the study	Nov. 2014 – Feb 2015				
1.1.4 Present study findings and recommendations including • partner/beneficiary clinics hospitals • needs assessment of facilities in two counties of Homa Bay and Kakamega	End Feb 2015				
1.2 Develop Project Budget based on inception report					?
1.3 Prepare advocacy materials/messages	March 2015		Onsite Project team		

1.4 Sensitise/orient County Health Management Teams and Leadership	March 2015	MOU or joint workplan signed with County /sub county public health officials and Shikuri Project			
1.5 Conduct community awareness in targeted counties.	March – June 2015		Project Coordinator /team		
1.6 Advocate for inclusion of SCD interventions in in County and Sub County District workplans for 2016-2017 planning cycle		District workplan which incorporates SCD for 2016			
Objective 2: Improve care of existing cases of SCD/SCA at County Level ?					
2.1 In two counties of Homa Bay and Kakamega, train health providers and update their knowledge and skills on SCD/SCA (initial focus on two sub-counties in each county)	March – June 2015				
2.2 As part of 2.1, prepare and distribute learning and caregiver/parental education materials to health workers on SCD/SCA	Feb – June 2015	Xxx			
2.3 Provide free and/or affordable medicines and essential supplies to targeted health facilities including: Penicillin prophylactics; Folic Acid; Pain medication; Hydration (ORS/ORT); Hydroxy Urea (to limit mortality and risk of strokes from low HGB); Antibiotics; Anti-Malarials; Thermometers	June-December 2015				
2.4 Improve supportive supervision to targeted sub-counties and health facilities	March – December 2015				
2.5 Monitor stock-outs of supplies and commodities in targeted sub-counties and health facilities	March – December 2015				
2.6 Introduce/pilot screening test for SCD and point of care test for anaemia. This may include collaboration with USA/NIH	June – Dec 2015		This may include collaboration		

			with USA/NIH-		
Objective 3: Improve community level awareness, syndromic diagnosis or screening of SCD and care/referral of cases.					
3.1 Prepare learning materials for community health workers on SCD/SCA.	March – June 2015				
3.2 Print and distribute SCD/SCA CHW learning materials.	June 2015				
3.3 Identify/develop appropriate screening package for use at level 1 (community), 2 and 3	March 2015				
3.4 Identify target Communities (est 1,000 under 18 patients) for piloting community level sickle cell disease activities in 4 sub counties (2 each in Kakamega, 2 Homa Bay) Identifying partner clinics/hospitals	June – Dec 2015				
3.5 Develop and disseminate communication messages for public education and awareness	March – Dec 2015				
3.6 Training/orient CHWs in the care (home care, early referral) of SCD/SCA.					
Objective 4: Support counties to undertake newborn screening as part of district health package at community (1), dispensary (2), and H levels (3)					
4.1 Share experiences from other countries	Feb – March 2015				
4.2 Develop policy guidelines on newborn screening within maternal-newborn care package	June – Dec 2015				
4.3 Adapt/develop newborn screening protocols	June Dec 2015				
4.4. Develop operational plan for screening newborn screening in two sub counties, with target of 1000 (anyone with family history,)	2016				
New Objective 6 : Infant diagnosis campaign conducted with Measles Vaccination Programme					
6.1 6.2 6.3					
Objective 5: Improved coordination of partners involved in care of SCD/SCA patients					
5.1. Map out all partners involved SCD/SCA in Kenya and other East African countries	Nov-February 2015				

5.2 Establish/strengthen coordination of partners –forming a stakeholders' forum orassociation	June 2015				
5.3 Produce SCD newsletter	June 2015				
5.4 Hold Annual SCD meetings	Annually				

XV. *Management Arrangements*

XVI. *Phase II*

ii. Incorporate Primary prevention: This includes screening and counselling of pregnant women. Genetic counselling and health promotion activities can lead to substantial reductions in the number of at-risk children.

iii. Based on results, lessons learned and evaluation of Phase I, expand programme to Coastal District and other underserved communities in E Africa and among Displaced Communities in Sub Saharan Africa.

XVII. *Potential Partners and Donors*

In addition to the Advisory Board and Partners identified in this document, the Project intends to enroll the participation of a number of other partners for technical, advocacy, and fund-raising efforts

XVIII. *Acknowledgements*

The President gratefully acknowledges the advice, encouragement and support of the currently active Advisory Board Members including the following:

- Dr William Macharia
- Dr Vincent Orinda
- Mrs Marjorie Newman-Williams
  - Independent Consultant
- Mrs Nardos Bekele Thomas, U N Resident Co-ordinator, UNDP, Kenya
- Mrs Rebeca de Los Rios, former Advisor, WHO
- Mr Hernan Rosenberg
- Ms Foulata Kwena
- Ms Nancy Yates
  Former Programme Advisor, UNDP

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Investigators: Jodi B Segal, MD, MPH, John J Strouse, MD, Mary Catherine Beach, MD, MPH, Carlton Haywood, MA, Catherine Witkop, MD, MPH, Haeseong Park, MD, MPH, Renee F Wilson, MSc, Eric B Bass, MD, MPH, and Sophie Lanzkron, MD.

Rockville (MD): Agency for Healthcare Research and Quality (US); February 2008.

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[i] **Hydroxyurea for the Treatment of Sickle Cell Disease**

Evidence Reports/Technology Assessments, No. 165

Investigators: Jodi B Segal, MD, MPH, John J Strouse, MD, Mary Catherine Beach, MD, MPH, Carlton Haywood, MA, Catherine Witkop, MD, MPH, Haeseong Park, MD, MPH, Renee F Wilson, MSc, Eric B Bass, MD, MPH, and Sophie Lanzkron, MD.

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