

Analyzing Needs and Potential Impacts of a Feasible Sickle Cell Screening Program within the current Sierra Leonean Healthcare System

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Abstract

Sickle cell anemia (SCA) is a common, life-threatening, but largely neglected inherited blood disorder. Estimates suggest that in some parts of sub-Saharan Africa, the probability of death among children with SCA could be as high as 50-90%. Although in high income countries, neonatal screening and early detection have been shown to significantly reduce these high mortality rates, SCA screening is not commonplace in sub-Saharan Africa due to the lack of feasible screening options and established intervention programs.

In order to address this need, previous work has focused on designing and optimizing a low-cost, easy-to-use, SCD diagnostic test to be used in Sierra Leone, and eventually throughout other low and middle income countries (LMICs). Additionally, fieldwork in Sierra Leone was conducted to learn about the current healthcare system and status of SCA.

In order to analyze the needs and potential impacts of a feasible sickle cell screening program within this current healthcare context, the current healthcare system was mapped and analyzed to determine potential areas for intervention. Additionally, a literature review was used to compare and analyze the feasibility of different screening and intervention program options within this context.

Although there are few SCA screening programs in sub-Saharan Africa to compare to, a literature review found that sickle cell screening could be cost effective, and highlighted several needs and potential challenges. Additionally, further analysis illustrated how Sierra Leone's healthcare system could offer unique opportunities in designing a screening program.

Introduction

Background

- Sickle cell disease (SCD) is a common [1], life-threatening [2], but largely neglected [3] inherited blood disorder.
- Globally, more than 300,000 people are born with SCD each year [2,4,5], with 85% of those babies being born in Africa [5].
- Although there is limited data available, estimates suggest that in some parts of sub-Saharan Africa, the probability of death among children with SCA could be as high as 50-90% [3], and could be responsible for up to 5%-16% of under-5 child mortality [6].
- Despite this high mortality, neonatal screening and early detection have been shown to significantly reduce mortality rates [1] by allowing for several potentially life-saving interventions, including penicillin prophylaxis [1,2], pneumococcal immunization [2], education on handling symptoms [1,2], and hydroxyurea treatments [2,5].
- The significant benefits of these early interventions has led SCD screening among newborns to become standard in the United States and other high-income countries for years [2,4], allowing mortality rates due to SCD to drop significantly [1].
- Despite these benefits, SCA screening is not commonplace in sub-Saharan Africa due to the lack of feasible screening options and established intervention programs [2].

Past Work

- In order to address this need, a team at Lehigh University has spent the last three years designing a low-cost, easy-to-use, sickle cell anemia diagnostic test to be used in Sierra Leone, and eventually throughout other low and middle income countries.
- These efforts have been focused on the optimization of materials and reagents and testing of different lateral flow device configurations in order to establish a design which meets the economic, usability, and reliability demands of a sickle cell screening device for use in LMICs.
- A proof-of-concept lateral flow device which utilizes a novel E-junction geometry in order to overcome the high concentration of hemoglobin in blood without requiring a dilution step has been designed
- Additionally, in the summer of 2019 fieldwork was conducted in Sierra Leone to learn about the healthcare system and current sickle cell screening and treatment programs

Methods

- The Sierra Leonean healthcare system was mapped based on fieldwork experience and analyzed for various potential points of intervention.
- Next, a literature review was conducted to compare and analyze different sickle cell screening and intervention program options currently being utilized in other LMICs.
- Additionally, statistical analysis was performed to compare and analyze the feasibility of screening and intervention programs
- And finally, the Sierra Leonean healthcare system was analyzed in comparison to other LMICs to better assess potential areas for intervention.

Results

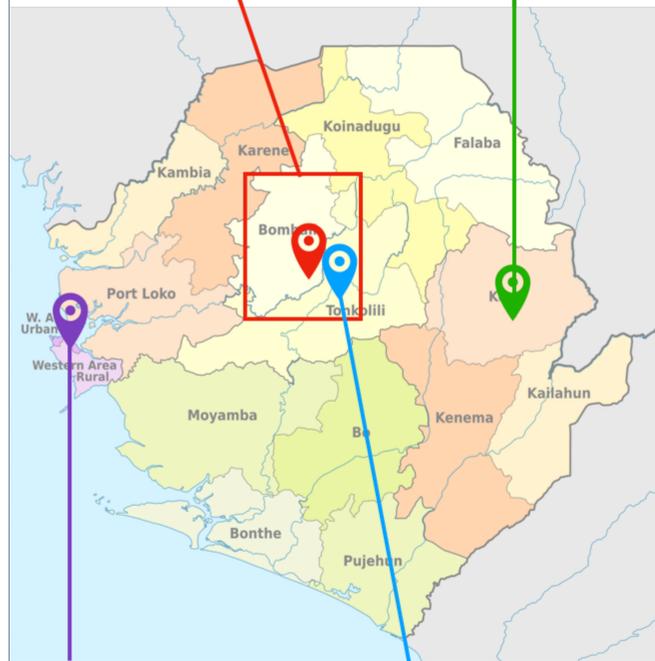
Sierra Leone: Current Sickle Cell Diagnosis and Intervention Options

Bombali & northern Tonkolili:

- Diagnosis
 - Child presents at CHC with symptoms
 - Referred to Makeni hospital where receive sickling test
- No treatment programs

Kono:

- Diagnosis
 - Child presents at CHC with symptoms
 - Referred to Koidu hospital where receive sickling test
- Intervention Program
 - Referred to Sickle Cell Carers Awareness Network (SCCAN)
 - Diagnosis confirmed with SickleSCAN
 - Receive counseling and education
 - Monthly clinic to receive vitamin C, folic acid, and penicillin prophylaxis



Freetown:

- Diagnosis
 - Child presents at hospital with symptoms
 - Sickling test performed
 - Sent to local lab for electrophoresis
- Intervention program
 - Referred to Sierra Leone Sickle Cell Society (SLSCS)
 - Receive genetic counseling and education
 - Monthly clinic to receive penicillin prophylaxis, folic acid, and malaria prophylaxis

Masanga Hospital

- NGO funded hospital with electrophoresis machine
- No specific screening or intervention program for sickle cell

Fig. 1. Summary of sickle cell screening and intervention options available throughout Sierra Leone based on fieldwork experience

Sierra Leone Healthcare System: Levels of Care

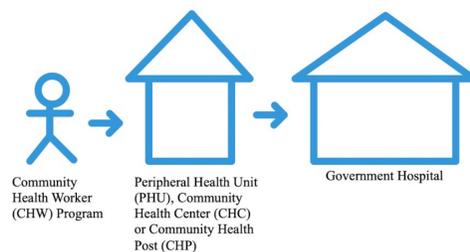


Fig. 2. Diagram of levels of care in Sierra Leone, highlighting the unique community health worker program which goes out into the community to refer people to come to their local health center

Results

Sierra Leone Healthcare System: Maternal and Child Health

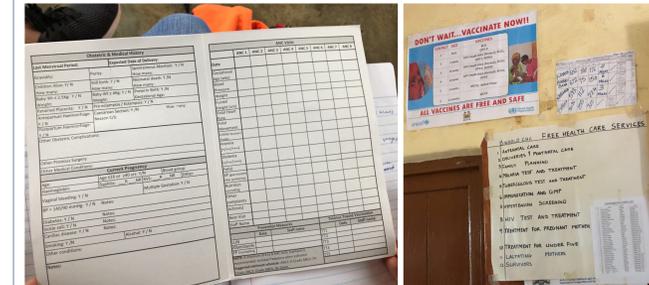
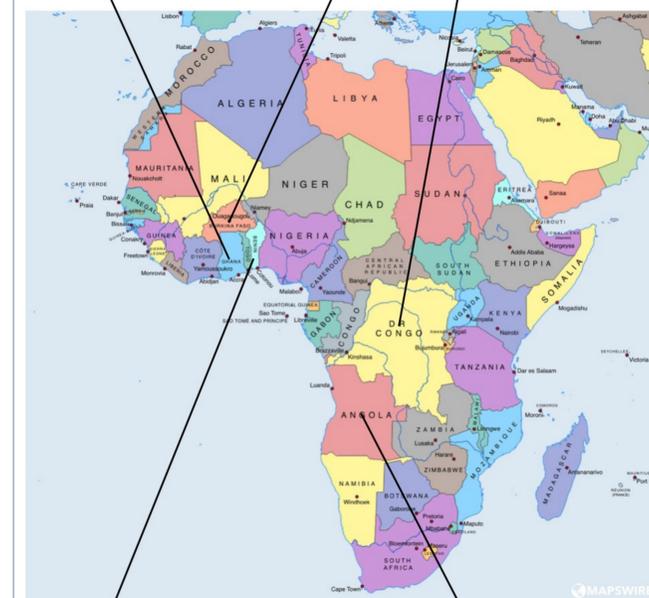


Fig. 3. Pictures showing examples of the maternal and child health services in Sierra Leone, where a screening cell program would likely be incorporated. (A) Picture of a "Maternity Record Card." The Sierra Leone government requires 4-8 antenatal care visits which are logged in this booklet. Additionally, women are legally required to give birth at a healthcare facility, with fines being imposed if you don't. (B) Picture of maternal and child health services offered a clinic. All clinics offer clinic days for pregnant mothers, children under 5, and for vaccines. Most women do bring their children for required checkups, however in some areas rainy season can prevent access due to flooding

Sickle Cell Screening and Intervention Programs in Other LMICs

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| Ghana [7,8]: <ul style="list-style-type: none"> Screening <ul style="list-style-type: none"> At the first well-baby visit 40 facilities in Kumasi and Accra plus two rural sites Central NBS laboratory performs testing with IEF Extensive tracking system Intervention <ul style="list-style-type: none"> Infant is placed on penicillin prophylaxis Comprehensive management is initiated at teaching hospital Genetic counseling | Burkina Faso [9]: <ul style="list-style-type: none"> Screening <ul style="list-style-type: none"> 5 public health centers in Ouagadougou Cord blood Commercially available IEF technique Confirmed using high-performance liquid chromatography (HPLC) in Belgium Interventions <ul style="list-style-type: none"> Information and education Malarial prophylaxis, penicillin prophylaxis, and pneumococcal vaccinations | Democratic Republic of the Congo [9]: <ul style="list-style-type: none"> Screening <ul style="list-style-type: none"> 17 health centers in Kinshasa Cord blood samples were collected on filter paper from all deliveries Screened and confirmed with Isoelectric focusing (IEF) Results confirmed at time of mandatory vaccinations Intervention <ul style="list-style-type: none"> Referred to specialist pediatrician Information and education Malarial prophylaxis, penicillin prophylaxis and pneumococcal vaccination |
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| Benin [10]: <ul style="list-style-type: none"> Screening <ul style="list-style-type: none"> At two of the largest maternity services in Cotonou Identified pregnant women at risk Mothers elect to have child tested Thin-layer agarose isoelectric focusing on dried blood collected Phenotypes were confirmed using citrate agar electrophoresis Interventions <ul style="list-style-type: none"> Enrolled into the comprehensive follow-up program when their mothers volunteered Anti-pneumococcal and anti-malarial prophylaxis, supplementation with folic and ascorbic acids, and specific immunizations During medical visits, parental education was continued, emphasizing the importance of keeping clinic appointments and regular follow-up | Angola [11, 12]: <ul style="list-style-type: none"> Screening <ul style="list-style-type: none"> Clinic at the National Children's Hospital in the capital city of Luanda Government mandated public health initiative Dried bloodspots from newborns Initially tested by IEF in the NBS laboratory at HPDB, the only dedicated pediatric hospital in Angola Samples with abnormal hemoglobin patterns were analyzed by capillary electrophoresis Families contacted by phone Intervention <ul style="list-style-type: none"> Enrolled in new born clinic Receive ongoing education and management Started on penicillin prophylaxis and given mosquito net and pneumococcal vaccine |
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Fig. 4. Summary of Sickle cell screening programs in other LMICs

Discussion

Big Picture in Sierra Leone

- Sickle cell is a largely neglected problem in Sierra Leone
- There are many misconceptions and lack of knowledge about sickle cell
- There are sickle cell programs in Freetown and Kono, however, there are no true diagnostic or treatment options available in most of the country
- The current healthcare system offers unique opportunities for implementing a screening program, such as with community health workers or during antenatal care visits
- Sickle cell programs in other LMICs and field work findings highlight several important considerations in designing a sickle cell screening program

Implications of Findings

- Epidemiological study is needed first
- Need confirmatory testing
- If tests sent away for collection, must ensure a way to give results to patients
- Intervention program must include education and genetic counseling in order to combat stigma and ensure compliance

Conclusions

- Sickle cell programs in other LMICs have proven that early screening and intervention is possible, and have proven to reduce the frequency and severity of sickle cell related acute complications
- Many challenges, such as overcoming stigma and ensuring follow ups, will need to be overcome
- Even with screening and intervention program, mortality will probably not be reduced to the same extent as in US

Future Prospects

- This project has outlined the current healthcare system in Sierra Leone, and identified potential areas for intervention based on current flow of care with maternal and child health services, as well as from published experiences with sickle cell screening programs in other LMICs
- Next steps involve using modeling and statistical analysis to further analyze feasible screening and intervention options; for example, comparing the feasibility, cost, and impact of a program with a cheaper, lower specificity test followed by shipping to a laboratory for isoelectric focusing (gold standard) versus a more expensive, higher specific point of care test
- In order to evaluate potential screening programs, epidemiologic data is needed to provide data on the prevalence and genotypes of sickle cell diseases in Sierra Leone

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