Background

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, maternal 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 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, a further analysis illustrated how Sierra Leone’s healthcare system could offer unique opportunities in designing a screening program.

Introduction

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% [6], and could be responsible for up to 5%-10% of under-5 child mortality [6].

Despite this high mortality, neonatal screening and early detection have been shown to significantly reduce mortality rates [7]. 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 [8]. However, maternal screening is not as common to detect family history, and thus reduce mortality rates due to SCD, to detect sickle cell trait [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 open 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 requiring a dilution step has been designed. Additionally, statistical analysis was performed to compare and analyze the feasibility of different screening and intervention programs.

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 and analyzes 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 Healthcare System: Maternal and Child Health

Sickle Cell Screening and Intervention Programs in Other LMICs

Sierra Leone: Current Sickle Cell Diagnosis and Intervention Options

Bombali & northern Tonkolili:

- Diagnosis
- Child presents at CHC with symptoms
- Routed to main hospital where receive hematological testing
- No confirmatory testing

Kono:

- Diagnosis
- Child presents at CHC with symptoms
- Routed to Koina Hospital where receive hematological testing
- No confirmatory testing

Intervention Program
- Routed to Sick Cell Care Awareness Network (SCKAN)
- Diagnose confirmed with SickcellSCAN
- Receive counseling and education

Monthly clinic to receive vitamins, folate, acid, and penicillin prophylaxis

Freetown:

- Diagnosis
- Child presents at hospital with symptoms
- Sickling not performed
- No confirmatory testing

Intervention program
- Routed in Sierra Leone Sickle Cell Society (SLSCS)
- Receive counseling and education

Monthly clinic to receive vitamins, folate, acid, and penicillin prophylaxis

Masanga Hospital:

- NO Sickle Cell screening and intervention program for sickle cell

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

Discussion

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 Room Card.” The Sierra Leone government requires 4-8 attended 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 bring their children for regular checkups, however in some areas many season can prevent access due to flooding

Fig. 4. Summary of Sickle cell screening programs in other LMICs

Results

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 LMIC’s 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 are sent for confirmatory, must ensure a way to get 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 LMIC’s 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 up, 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 creating 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 screening to a laboratory for nucleic acid testing (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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References


