Uganda Sickle Survey Results and Neonatal Screening Program

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Study Questions

1. What is the prevalence of sickle cell trait and disease in Uganda, by region and district?

2. What is the distribution of sickle cell trait across the country?

3. What is the relationship between sickle cell trait and malaria prevalence?

4. What is the early mortality of sickle cell disease?

5. What is the co-morbidity of HIV and sickle cell disease?
## Sickle Cell Trait: Prevalence by Region

<table>
<thead>
<tr>
<th>Region</th>
<th>Normal</th>
<th>Variant</th>
<th>Trait</th>
<th>Trait (%)</th>
<th>Disease</th>
<th>Disease (%)</th>
<th>TOTAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Central 1</td>
<td>12187</td>
<td>30</td>
<td>1802</td>
<td>12.80</td>
<td>64</td>
<td>0.45</td>
<td>14083</td>
</tr>
<tr>
<td>Central 2</td>
<td>9048</td>
<td>31</td>
<td>1486</td>
<td>13.95</td>
<td>85</td>
<td>0.80</td>
<td>10650</td>
</tr>
<tr>
<td>East Central</td>
<td>4911</td>
<td>42</td>
<td>1201</td>
<td>19.24</td>
<td>89</td>
<td>1.43</td>
<td>6243</td>
</tr>
<tr>
<td>Kampala</td>
<td>11006</td>
<td>40</td>
<td>1725</td>
<td>13.42</td>
<td>81</td>
<td>0.63</td>
<td>12852</td>
</tr>
<tr>
<td>Mid Eastern</td>
<td>3835</td>
<td>35</td>
<td>721</td>
<td>15.52</td>
<td>55</td>
<td>1.18</td>
<td>4646</td>
</tr>
<tr>
<td>Mid Northern</td>
<td>9681</td>
<td>121</td>
<td>2359</td>
<td>19.16</td>
<td>153</td>
<td>1.24</td>
<td>12314</td>
</tr>
<tr>
<td>Mid Western</td>
<td>10897</td>
<td>31</td>
<td>1358</td>
<td>11.00</td>
<td>59</td>
<td>0.48</td>
<td>12345</td>
</tr>
<tr>
<td>North East</td>
<td>3517</td>
<td>30</td>
<td>651</td>
<td>15.36</td>
<td>40</td>
<td>0.94</td>
<td>4238</td>
</tr>
<tr>
<td>South Western</td>
<td>12440</td>
<td>16</td>
<td>586</td>
<td>4.48</td>
<td>25</td>
<td>0.19</td>
<td>13067</td>
</tr>
<tr>
<td>West Nile</td>
<td>2421</td>
<td>50</td>
<td>394</td>
<td>13.69</td>
<td>13</td>
<td>0.45</td>
<td>2878</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>79943</td>
<td>426</td>
<td>12283</td>
<td><strong>13.16</strong></td>
<td>664</td>
<td><strong>0.71</strong></td>
<td><strong>93316</strong></td>
</tr>
</tbody>
</table>
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Uganda Regional Map (US3 Data)
Sickle Cell Trait

Mid Northern
MP 63%

North East
MP 40%

Sickle Cell Trait
19.2%

Sickle Cell Trait
13.7%
Highest Prevalence Districts

49 of the 112 districts have sickle cell trait >15.0%
8 districts have sickle cell trait >20.0%
   - Bundibugyo 21.9%, Bulisa 22.1%, Alebtong 24.3%
   - Jinja 18.9%, Tororo 19.5%, Gulu 19.6%, Lira 20.0%

14 districts contain 47% of the sickle cell disease
   - Kampala, Gulu, Lira, Jinja, Tororo, Luweero, Wakiso
   - Apac, Iganga, Muyuge, Buikwe, Oyam, Masaka, Masindi

Screening should focus on highest burden regions
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Uganda Regional Maps (US3 Data)
Sickle Cell Trait and Malaria
Malaria Parasites and Sickle Cell Trait

Malaria Parasites (% with + microscopy) vs. Sickle Cell Trait (%)

R = 0.69
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**Sickle Cell Disease: Early Mortality**

EID samples are between birth and 18 months of age.

Analysis of sickle cell trait and disease by age.

Predictions:
- Rate of sickle cell trait will increase (malaria survival)
- Rate of sickle cell disease will decrease (mortality)

<table>
<thead>
<tr>
<th>Age Range</th>
<th>Normal (%)</th>
<th>Trait (%)</th>
<th>Disease (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth – 6.0 months</td>
<td>85.63</td>
<td>13.09</td>
<td>0.76</td>
</tr>
<tr>
<td>6.1 – 12.0 months</td>
<td>85.78</td>
<td>13.27</td>
<td>0.64</td>
</tr>
<tr>
<td>12.1 – 18.0 months</td>
<td>85.73</td>
<td>13.36</td>
<td>0.57</td>
</tr>
</tbody>
</table>

*25% Mortality*
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Sickle Cell Disease and HIV

All EID samples are from HIV-exposed infants

~5% of the EID samples are HIV-positive

Prediction:

In (+) HIV infants, sickle cell disease is associated with early death

<table>
<thead>
<tr>
<th></th>
<th>Normal</th>
<th>Trait</th>
<th>Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>HIV Negative Infants</td>
<td>86.1%</td>
<td>13.2%</td>
<td>0.73%</td>
</tr>
<tr>
<td>HIV Positive Infants</td>
<td>86.6%</td>
<td>13.0%</td>
<td>0.43%</td>
</tr>
</tbody>
</table>

41% Mortality
Opportunities to Neonatal Sickle Cell Screening

• Results of the sickle cell prevalence study
• Laboratory capacity built through the study
• Sample and results transport network
Early Diagnosis

CPHL Sickle Cell Laboratory with Annual Testing Capacity of >600,000 tests
The Hub-based National Specimens and Result Transportation Network

Structure of the hub network

Map showing current Hub Distribution

The bike and rider given to each hub/district, Holly Foundation clinic

- 82 hubs reaching 2400 health facilities with viable laboratories conducting most of the tests for the 30 or so lower facilities in its catchment
- Strategy is to have 100 hubs and strengthen lab services such that lower sites access them thru the NSRTN
<table>
<thead>
<tr>
<th>District</th>
<th>No. of sites</th>
<th>No. tested</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gulu</td>
<td>42</td>
<td>2158</td>
</tr>
<tr>
<td>Lira</td>
<td>31</td>
<td>1781</td>
</tr>
<tr>
<td>Kitgum</td>
<td>19</td>
<td>963</td>
</tr>
<tr>
<td>Oyam</td>
<td>33</td>
<td>1451</td>
</tr>
<tr>
<td>Dokolo</td>
<td>16</td>
<td>67</td>
</tr>
<tr>
<td>Jinja</td>
<td>28</td>
<td>1936</td>
</tr>
<tr>
<td>Tororo</td>
<td>41</td>
<td>2031</td>
</tr>
<tr>
<td>Makerere launch</td>
<td></td>
<td>1803</td>
</tr>
<tr>
<td>Eden Church</td>
<td></td>
<td>237</td>
</tr>
<tr>
<td>Namasuba College</td>
<td></td>
<td>103</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>211</strong></td>
<td><strong>12,530</strong></td>
</tr>
</tbody>
</table>
Care and Treatment

Early care and treatment
- Penicillin prophylaxis
- Malaria control and prophylaxis
- PCV (pneumococcal) and other immunizations
- Clinical management
- Health education
- Screening for stroke risk
- Home management

Enroll the baby into sickle cell care
- Dedicated sickle cell clinic days and staff
- Establish routine follow-up
- Establish emergency SCD care plan
- Continued family education
Acknowledgements

• Cincinnati Children’s Hospital in the USA
• Dedication and support of the CPHL leadership and laboratory staff
• Makerere University for support of research and educational efforts
• MoH for vision and planning to allow this project to move forward