ASH, in partnership with the United States Department of Health and Human Services and SickleinAfrica Presents:

Addressing the Global Burden of Sickle Cell Disease During the COVID-19 Pandemic and Beyond

Monday, June 29, 2020
Introduction To Webinar

Martha Liggett, Esq
Executive Director, American Society of Hematology
Opening Remarks

Dr. Julie Makani
Associate Professor and Head of the Department of Hematology and Blood Transfusion at Muhimbili University of Health and Allied Sciences

Dr. Alexis Thompson
Section Chief of Hematology in the Department of Pediatrics Northwestern University School of Medicine
Sickle Cell Disease and COVID19 in Africa

June 2020
Sickle Cell Disease

Social and Cultural factors

Environmental factors
Sickle Cell Disease and COVID-19 in Africa

- **SCD**
  - Global SCD: 5,000,000*
  - SCD in Africa: 3,500,000*
  - Proportion in Africa: 75%

- **SCD and COVID-19**
  - Testing and Treatment
  - COVID-19 on SCD
  - COVID-19 and SCD

- **COVID-19**
  - Global COVID19 Cases: 10,000,000*
  - COVID19 in Africa: 350,000*
  - Proportion in Africa: 5%

**CHARTA:** Consortium of Health, Advocacy, Research and Training in Africa
Thank You! Asante!

Individuals, Community, Healthcare Providers, Researchers, Government, Industry, Non-government Organizations, Funders

SickleInAfrica is supported by the National Heart, Lung, And Blood Institute of the US National Institutes of Health under Award Number U24HL135881 – Sickle Pan-African Research Consortium (SPARCo).

The content is solely the responsibility of the authors and does not necessarily represent the official views of the National Institutes of Health.
Welcoming Remarks

Admiral Brett Giroir
Assistant Secretary of Health, United States
Department of Health and Human Services
Welcoming Remarks

Pr. Jean-Marie Dangou
Coordinator, Noncommunicable Diseases
Management World Health Organization Regional Office for Africa
Welcoming Remarks

Bukky Bolarinwa
President, Sickle Cell Aid Foundation
COVID-19 and Sickle Cell Disease Presentations
Blood Transfusion for SCD in Africa: Impact of COVID-19

Isaac Odame
Hospital for Sick Children & University of Toronto
Global Sickle Cell Disease Network
Evidence-Based Management of Sickle Cell Disease

Expert Panel Report, 2014


JAMA 2014;312(10):1033-1048
<table>
<thead>
<tr>
<th>Complication</th>
<th>Transfusion Method</th>
<th>Strength of Recommendation</th>
<th>Quality of Evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptomatic severe acute chest syndrome (O₂ sats &lt; 90% despite supplemental O₂)</td>
<td>Exchange</td>
<td>Strong</td>
<td>Low</td>
</tr>
<tr>
<td>Acute splenic sequestration with severe anemia</td>
<td>Simple</td>
<td>Strong</td>
<td>Low</td>
</tr>
<tr>
<td>In children and adults with acute stroke, initiate monthly transfusions</td>
<td>Simple or exchange</td>
<td>Moderate</td>
<td>Low</td>
</tr>
<tr>
<td>Hepatic sequestration</td>
<td>Exchange or simple</td>
<td>Consensus</td>
<td>Panel</td>
</tr>
<tr>
<td>Intrahepatic cholestasis</td>
<td>Exchange or simple</td>
<td>Consensus</td>
<td>Panel</td>
</tr>
<tr>
<td>Multisystem organ failure</td>
<td>Exchange or simple</td>
<td>Consensus</td>
<td>Panel</td>
</tr>
<tr>
<td>Aplastic crisis</td>
<td>Simple</td>
<td>Consensus</td>
<td>Panel</td>
</tr>
<tr>
<td>Symptomatic anemia</td>
<td>Simple</td>
<td>Consensus</td>
<td>Panel</td>
</tr>
<tr>
<td>Child with TCD reading &gt; 200 cm/s</td>
<td>Exchange or simple</td>
<td>Strong</td>
<td>High</td>
</tr>
<tr>
<td>Adults and children with previous clinically overt stroke</td>
<td>Exchange or simple</td>
<td>Moderate</td>
<td>Low</td>
</tr>
<tr>
<td>Age</td>
<td>All patients (n=83)</td>
<td>Patients aged 0-14 years (n=12)</td>
<td>Patients aged 15-44 years (n=56)</td>
</tr>
<tr>
<td>---------------------------</td>
<td>---------------------</td>
<td>----------------------------------</td>
<td>----------------------------------</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>38 (46)</td>
<td>6 (50)</td>
<td>22 (39)</td>
</tr>
<tr>
<td>Female</td>
<td>45 (54)</td>
<td>6 (50)</td>
<td>34 (61)</td>
</tr>
<tr>
<td>Haemoglobin genotype</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>SS/SS^o</td>
<td>71 (86)</td>
<td>11 (92)</td>
<td>48 (86)</td>
</tr>
<tr>
<td>SC</td>
<td>8 (10)</td>
<td>0</td>
<td>5 (9)</td>
</tr>
<tr>
<td>SB^+</td>
<td>4 (5)</td>
<td>1 (8)</td>
<td>3 (5)</td>
</tr>
<tr>
<td>Hydroxyurea treatment at admission</td>
<td>38 (46)</td>
<td>4 (33)</td>
<td>28 (50)</td>
</tr>
<tr>
<td>Hydroxyurea dose (mg/kg/day)</td>
<td>17.9 (8.8-30.2)</td>
<td>18.8 (18.6-23.3)</td>
<td>18.2 (11.8-30.2)</td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>68 (54-110)</td>
<td>32 (5-49)</td>
<td>171 (41-110)</td>
</tr>
<tr>
<td>Vaso-occlusive crisis</td>
<td>44/81* (54)</td>
<td>6 (50)</td>
<td>34 (61)</td>
</tr>
<tr>
<td>Acute chest syndrome</td>
<td>23/82* (28)</td>
<td>2 (17)</td>
<td>17 (30)</td>
</tr>
<tr>
<td>Transfusion</td>
<td>31 (37)</td>
<td>4 (33)</td>
<td>18 (32)</td>
</tr>
<tr>
<td>Length of hospital stay (days)</td>
<td>8 (2-37)</td>
<td>4 (2-10)</td>
<td>7 (2-35)</td>
</tr>
<tr>
<td>Mechanical ventilation in the intensive care unit</td>
<td>9/17 (53)</td>
<td>0</td>
<td>3/7 (43)</td>
</tr>
</tbody>
</table>

Data are n (%), n/N (%), or median (range). Percentages do not always equal 100% because of rounding. Ethnicity data were not collected in line with usual practice in France.

*Data for vaso-occlusive crisis were not available for two patients and acute chest syndrome not available for one patient. †Simple transfusion or exchange transfusion (manual or automated) during the hospital stay. ‡Hospitalisation was completed for 80 (96%) of 83 patients and is ongoing at the date of the notification for the other three. §17 patients were admitted to the intensive care unit.

Jean-Benoit A et al. Lancet Haematol, June 18, 2020

https://doi.org/10.1016/S2552-3026

Table 1: Patient characteristics by age range
RBC Demand Far Exceeds Supply in Africa

Lund TC et al. Transf Apheres Sci (2013), http://dx.doi.org/10.1016/j.transci.2013.06.014
WHO estimates that 10 donations per 1000 population are necessary to sustain any country’s blood transfusion requirements.
The impact of COVID-19 on blood supply: Ghana

• Cancellation of mobile outreaches for mass blood donation events.
  • 90% of voluntary blood donations - mobile outreaches; only 10% are walk-in
• Closure of institutions: schools, churches, mosques, corporate organizations
• Reduced individual blood donations at fixed sites
  • Loss of replacement donors on account of cancellation of elective surgeries
  • Restrictions in movement
  • Fear of visiting hospital-based collection sites
• Reduced group blood donations at fixed sites: restrictions in public gatherings
• Reduced prioritisation of blood donation
  • Public anxiety
  • Economic stress accompanying sudden lockdown
The impact of COVID-19 on blood supply

• Voluntary donations
  • more severely impacted than replacement donations.

• Adverse affects availability of blood for emergency transfusions.
Emergency Response Plan

Goals of plan
1. Maintain enough stocks of blood and components for emergency transfusions
2. Minimize risk of community spread of COVID-19 through blood donation activities

Support for the plan obtained from:
- Ministry of Health and National COVID-19 Task Force
- Hospitals and facilities involved in blood collection
- Media organisations

Key areas targeted: blood donor recruitment
blood collection (donor care)
hospital supply
Public Awareness Campaign

• Key Messages
  • The continuing need for blood donation
  • Measures taken by Blood Centres to prevent coronavirus spread
  • Request to contact blood service for blood donation appointments
  • Self-deferral for potential donors with possible COVID-19 exposure or symptoms

• Communication Channels
  • Local radio and TV, newspapers
  • Social media platforms
  • Bulk text messaging/direct phone calls to blood donors and representatives of blood donor groups
Donor Recruitment

• Accessible and spacious sites to operate fixed blood donation clinics and mobile blood collection trailer

• Contact lists of blood donors for systematic tele-recruitment

• Centrally-coordinated Exemption Pass issued to blood donors

• ‘Thank you’ text messages sent to all donors within 24 hours.

• Ongoing survey of donors attending donor clinics: effectiveness of the communication channels
Donor Selection

• Temperature checks using non-contact thermometers

• Questionnaire assessment of potential donors for COVID-19 exposure or symptoms

• Only potential donors who passed COVID-19 pre-donation assessment proceed to standard pre-donation assessment
Success and Challenges

• Noticeable response in both walk-in donors and mobile outreaches

• Overall, blood supply sourced from a combination of both voluntary and replacement donations

Challenges

• Cost-effectiveness of low-yielding mobile outreaches

• Only 33% of blood requests supplied during the period.
Hydroxyurea therapy and need for RBC transfusion in Africa

NOHARM

Robert O. Opoka et al. Blood 2017;130:2585-2593

REACH

## NOHARM-MTD Trial Results


<table>
<thead>
<tr>
<th>Event</th>
<th>Fixed-Dose Group (N = 94)</th>
<th>Dose-Escalation Group (N = 93)</th>
<th>Incidence Rate Ratio in Dose-Escalation Group (95% CI)</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Serious adverse events</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sickle cell–related</td>
<td>6</td>
<td>5</td>
<td>0.84 (0.24–2.79)</td>
<td>0.77</td>
</tr>
<tr>
<td>Non–sickle cell–related</td>
<td>1</td>
<td>0</td>
<td>---</td>
<td>---</td>
</tr>
<tr>
<td><strong>Clinical adverse events</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sickle cell–related</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Any grade</td>
<td>245</td>
<td>105</td>
<td>0.43 (0.34–0.54)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Grade ≥3</td>
<td>136</td>
<td>48</td>
<td>0.36 (0.25–0.49)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Non–sickle cell–related</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Any grade</td>
<td>321</td>
<td>205</td>
<td>0.64 (0.54–0.77)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Grade ≥3</td>
<td>93</td>
<td>54</td>
<td>0.59 (0.42–0.82)</td>
<td>0.002</td>
</tr>
<tr>
<td>Malaria infections</td>
<td>6</td>
<td>3</td>
<td>0.50 (0.11–1.91)</td>
<td>0.33</td>
</tr>
<tr>
<td><strong>Clinical complications of sickle cell anemia</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vaso-occlusive pain</td>
<td>200</td>
<td>86</td>
<td>0.43 (0.34–0.56)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Acute chest syndrome or pneumonia</td>
<td>30</td>
<td>8</td>
<td>0.27 (0.11–0.56)</td>
<td>0.001</td>
</tr>
<tr>
<td>Acute splenic sequestration</td>
<td>14</td>
<td>8</td>
<td>0.58 (0.23–1.34)</td>
<td>0.21</td>
</tr>
<tr>
<td>Stroke or transient ischemic attack</td>
<td>0</td>
<td>0</td>
<td>---</td>
<td>---</td>
</tr>
<tr>
<td><strong>Clinical interventions</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Transfusions</td>
<td>116</td>
<td>34</td>
<td>0.30 (0.20–0.43)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Hospitalizations</td>
<td>90</td>
<td>19</td>
<td>0.21 (0.13–0.34)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td><strong>Laboratory dose-limiting toxic effects</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Anemia</td>
<td>12</td>
<td>9</td>
<td>0.76 (0.31–1.79)</td>
<td>0.33</td>
</tr>
<tr>
<td>Reticulocytopenia</td>
<td>13</td>
<td>13</td>
<td>1.01 (0.46–2.20)</td>
<td>0.98</td>
</tr>
<tr>
<td>Neutropenia</td>
<td>4</td>
<td>8</td>
<td>2.02 (0.64–7.56)</td>
<td>0.25</td>
</tr>
<tr>
<td>Thrombocytopenia</td>
<td>14</td>
<td>13</td>
<td>0.94 (0.43–2.00)</td>
<td>0.86</td>
</tr>
</tbody>
</table>

*Clinical adverse events do not include serious adverse events or laboratory adverse events. Incidence rate ratios with 95% confidence intervals (CIs) were calculated according to follow-up time.

† Anemia was defined as a hemoglobin level of less than 4 g per deciliter, reticulocytopenia as an absolute reticulocyte count of less than 80x10⁹ per liter, neutropenia as an absolute neutrophil count of less than 1.0x10⁹ per liter, and thrombocytopenia as a platelet count of less than 1.0x10⁹ per liter.
How to improve access to blood transfusion for SCD in LICs

• Increase funding for blood transfusion services
• Increase blood donations
  • Voluntary non-remunerated donors VS
  • Family/replacement donors
• Improve the utilisation of available units
• Widespread use of hydroxyurea in patients with SCD
• Innovative technology to deliver blood where most needed
Drones to deliver blood

• Started in Rwanda in Oct 2016
  Speed of 100 km/hour
  Distance of 74 km
  Delivered > 2600 units of blood

• Similar service in Ghana since April 2019

https://www.wired.com/story/africas-delivery-drones-are-zipping-past-the-us/
Acknowledgements

• Dr. Justina Ansah
  Director, National Blood Service Ghana

• Prof. Heather Hume
  Department of Paediatrics, University of Montreal, Canada
  Department of Paediatrics and Child Health, Makerere University, Uganda

Thank You
Sustaining Service Delivery for Sickle Cell Care in Uganda amidst COVID-19

COVID-19 and SCD Coalition Webinar Discussion

June 29, 2020

Dr. Charles Kiyaga,
National Sickle Cell Program Manager,
MINISTRY OF HEALTH, UGANDA.
Email: ckiyaga@gmail.com
Sickle is one of the major causes of child mortality. It is estimated that world over 300,000 to 400,000 babies are born each with sickle cell disease, 75% of whom are born in SSA, but 70% to 80% never live to sickle their 5th birth day.

In Uganda it is estimated that 17,000 to 20,000 babies are born each year with SCD, but about 80% never live to see their 5th birth day. Sickle cell is responsible for about 18% of under 5 mortality in Uganda. However the full extent of the burden was never known until we undertook a nationwide survey code named US3, in 2014 - 2015.

It was a One year cross-sectional study where close to 100,000 HIV exposed infants from the EID program were tested.

Isoelectric Focusing (IEF) was the technology used on DBS samples.
Results of the Uganda Sickle Surveillance study

- ~100,000 samples were considered
- 13.3% were Sickle cell traits
- 0.7% had Sickle cell disease
- 49 of the 112 districts have sickle cell trait >15.0%
- 8 districts have sickle cell trait >20.0%
- 14 districts account for 47% of the national burden
Strategies Following the Survey

The survey results were an eye-opener and led to several strategies;

- Initiation of targeted Newborn screening in high burden districts
- Initiation of sickle cell clinics in major hospitals in high burden districts, and capacity building for health care workers
- Establishment of National sickle cell NBS laboratory and a mechanism for sample and results transport
- Standardization of the management of sickle cell disease
- Establishment of strategic partnerships with Cultural, religious and political leaders
- Coordinating NGOs in the space of SCD through the NGO forum.
- Forming a national sickle cell steering committee
- Forming a committee of senior clinicians to support the clinics
- Working closely with the media for sickle cell sensitisation
Onset of COVID-19
Circumstances around COVID-19

The Coronavirus disease 2019 (COVID-19) caused by Coronavirus-2 (SARS-CoV-2) started as an outbreak in China in December 2019

On 11\textsuperscript{th} March 2020:
- Over 114 countries affected
- More than 118,000 cases
- More than 4,291 deaths

Hence WHO declared COVID-19 a pandemic, calling upon all countries to quickly put up control measures

Following this, Uganda instituted a national lock down restricting movement of people, both private and public transport and closing of most businesses, retaining only essential services.

These measures affected delivery of health care; including SCD care.
Challenges in SCD care due to COVID-19

With COVID and the Lockdown;

- SCD patients are at high risk of developing complications if they contract COVID-19: due to their vulnerable health condition
- SCD patients could no longer move to the hospital with ease: many were developing complications or even dying
- Economic hardships made survival and care more challenging: Lack of money to buy both drugs and food for SCD patients in families
- Increased anxiety, stress and depression leading to increased occurrences of SCD crises
- Continued shift of attention from other diseases SCD inclusive to only COVID-19.
- Limited data on the effects of COVID-19 on SC patients in Uganda.
- Limited funding to mitigate the challenges caused by COVID-19 in SCD care

MOH and the National SCD steering committee convened to design strategies to ensure continuity of provision of SCD care
Devised Solutions to the Challenges

- Continuing lobbying and advocacy to attend to the needs of the SCD program despite the COVID-19 pandemic
- Hiring transportation for health workers. Public and private transport has now been eased
- Increased advocacy with Government and partners to increase funding for the SCD program to mitigate the challenges caused by COVID-19
- Continuous awareness and sensitization of SCD clients about ways to prevent contacting COVID-19
- The use of social media to pass communication on to sickle cell patients and their families
- Increased counselling and provision of psychosocial support to avert the dangers of anxiety and depression among SCD clients
- Planning to undertake a qualitative study to know the effects of COVID-19 on SCD
Continuity of Newborn Sickle Cell Screening

Private vehicles were hired to assist in transportation of laboratory workers to ensure continuity of newborn screening.

Midwives and all health facilities were notified that Newborn screening was still taking place and they were encouraged to continue sending samples.

The newborn screening laboratory has continued to run samples 5 days a week.
The SCD program has over the time created over 35 sickle cell clinics across the country to provide routine SCD care. All these clinics were notified about the rationale of continuing SCD care amidst the COVID-19 pandemic.

The Department of NCDs in MOH also lobbed with Government to ensure that patients with NCDs; including SCD requiring medical care should be allowed to move to health facilities, amidst the Lockdown.
Use of Social media to engage and sensitize SCD patients

- Working with NGOs and patient networks, the national SC program created several social media platforms, to reach out to people living with SCD. These platforms have been used to sensitize them about COVID-19 and how they can still continue to get the necessary health care that they need.

- Through social media, health workers are offering free consultation and medical advice to different SCD patients who might be unable to move to the hospital.
Commemoration of the World SCD Day on June 19, 2020

The program has been commemorating the World SCD day on 19th June of ever year since 2015; gathering between 2,000 to 3,000 people and stake holders at each.

COVID-19 and the Lockdown affected this year’s commemoration and hence a scientific commemoration involving 30 key stake holders was convened this year. This function was broadcasted live on the national TV and also on Facebook Live and Zoom.

Government and other stake holders were mobilized and encouraged to support SCD care despite the current crisis of COVID-19.
National SCD management and prevention guidelines launched

- The National SCD management and prevention guidelines were launched on this year’s World SCD day commemoration, to standardize sickle cell care at all levels.

- A partnership with Novartis was launched aimed at increasing access to Hydroxyurea at affordable cost, and also to manufacture pediatric formulation.

- Our theme this year is to advance case for sickle cell disease and to get every body involved.
Acknowledgements

- We appreciate our collaborating partners Cincinnati Children’s Hospital based in the US for the technical and logistical support.

- Thanks to Perkin Elmer for the logistical support.

- Thanks to Novartis for supporting the World SCD day.

- We acknowledge Makerere University for supporting the study.

- We thank all health workers and stakeholders for the usual cooperation which enables the program activities to continue.
Thanks For Listening
Addressing the Global Burden of Sickle Cell Disease During the COVID-19 Pandemic and Beyond

Sickle Cell Disease with COVID-19 Registries

Prof. Kwaku Ohene-Frempong, MD
Sickle Cell Foundation of Ghana
Clinical Characteristics of Coronavirus Disease 2019 in China

COVID-19: Clinical Characteristics (2)

Table 1. Clinical Characteristics of the Study Patients, According to Disease Severity and the Presence or Absence of the Primary Composite End Point.*

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>All Patients (N = 1099)</th>
<th>Disease Severity</th>
<th>Presence or Absence of the Primary Composite End Point.*</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Non-severe (N = 926)</td>
<td>Severe (N = 173)</td>
</tr>
<tr>
<td>Symptoms</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cough</td>
<td>745 (67.8)</td>
<td>623 (67.3)</td>
<td>122 (70.5)</td>
</tr>
<tr>
<td>Fatigue</td>
<td>419 (38.1)</td>
<td>350 (37.8)</td>
<td>69 (39.9)</td>
</tr>
<tr>
<td>Sputum production</td>
<td>370 (33.7)</td>
<td>309 (33.4)</td>
<td>61 (35.3)</td>
</tr>
<tr>
<td>Shortness of breath</td>
<td>205 (18.7)</td>
<td>140 (15.1)</td>
<td>65 (37.6)</td>
</tr>
<tr>
<td>Myalgia or arthralgia</td>
<td>164 (14.9)</td>
<td>134 (14.5)</td>
<td>30 (17.3)</td>
</tr>
<tr>
<td>Sore throat</td>
<td>153 (13.9)</td>
<td>130 (14.0)</td>
<td>23 (13.3)</td>
</tr>
<tr>
<td>Headache</td>
<td>150 (13.6)</td>
<td>124 (13.4)</td>
<td>26 (15.0)</td>
</tr>
<tr>
<td>Chills</td>
<td>126 (11.5)</td>
<td>100 (10.8)</td>
<td>26 (15.0)</td>
</tr>
</tbody>
</table>

The primary composite end point was admission to an intensive care unit, the use of mechanical ventilation, or death.

Guan et al., 2020
Global Burden of Sickle Cell Disease During the COVID-19


Patients with no chronic renal, heart, or debilitating stroke

<table>
<thead>
<tr>
<th>Circumstance</th>
<th>No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain, uncomplicated</td>
<td>25</td>
</tr>
<tr>
<td>Pain with acute chest syndrome (ACS)</td>
<td>20</td>
</tr>
<tr>
<td>ACS</td>
<td>9</td>
</tr>
<tr>
<td>Stroke</td>
<td>15</td>
</tr>
<tr>
<td>Peri-operative</td>
<td>14</td>
</tr>
<tr>
<td>Infection</td>
<td>13</td>
</tr>
<tr>
<td>Cancer</td>
<td>4</td>
</tr>
<tr>
<td>Gastrointestinal bleeding</td>
<td>3</td>
</tr>
<tr>
<td>Trauma</td>
<td>14</td>
</tr>
<tr>
<td>Suddenly at home</td>
<td>10</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>7</td>
</tr>
<tr>
<td>Circumstances not known</td>
<td>37</td>
</tr>
</tbody>
</table>
COVID-19 and SCD: The Need for Registries

• To pool clinical information as quickly as possible in order to collect data to support clinical management

• To quickly detect opportunities for clinical research whose outcomes may improve clinical management

• To determine unique features of COVOD-19 in SCD population
COVID-19 and SCD: Registries Data not Adjudicated

• Clinical phenotypes not strictly defined

• Laboratory methods and imaging techniques not uniform

• Observations not designed to meet clinical trial quality
COVID-19 and SCD: Registries and Surveys

1. USA:
   • Secure SCD
   • American Society of Hematology
     (In development)

2. UK -

3. France -
Global Burden of Sickle Cell Disease During the COVID-19

COVID-19 and SCD: Registries and Surveys

ASH RC COVID-19 Registry for Hematology

The ASH Research Collaborative (ASH RC) COVID-19 Registry for Hematology, a global public reference tool that is part of the ASH RC Data Hub platform, captures data on individuals who test positive for COVID-19 and have a hematologic condition (past or present) and/or have experienced a post-COVID-19 hematologic complication. As data are received and analyzed, real-time observational summaries are made available via a dashboard.

[Enter a Case] [Data Summaries]
Real-time national survey of COVID-19 in haemoglobinopathy and rare anaemia patients

Dr Paul Telfer
Centre for Genomics and Child Health
Blizard Institute, Queen Mary University of London
Royal London Hospital, Barts Health NHS Trust, London, UK

Date: Sunday 14th June
Program section: Late breaking oral session
Aim of survey

• To provide national data to guide patient management during the COVID19 epidemic
Accrual
Conclusions

- Most COVID19 cases have been clinically mild
- Relatively few children have been infected, none have required respiratory support or have died
- Increasing age is a risk factor for worse outcome
- Although not statistically significant, milder genotypic severity showed a trend to increased severity
- Men were not more likely to have a worse outcome compared to women
On March 13, 2020, at an early stage of the COVID-19 pandemic in France, we invited all practitioners involved in the management of patients with sickle cell disease to report on all inpatients with sickle cell disease and confirmed COVID-19 by RNA.
## COVID-19 and SCD – French Experience

<table>
<thead>
<tr>
<th></th>
<th>All patients (n=83)</th>
<th>Patients aged 0-14 years (n=12)</th>
<th>Patients aged 15-44 years (n=56)</th>
<th>Patients aged 45-64 years (n=14)</th>
<th>Patients aged 65-74 years (n=1)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age</strong></td>
<td>30 (0-3-68)</td>
<td>--</td>
<td>--</td>
<td>--</td>
<td>--</td>
</tr>
<tr>
<td><strong>Sex</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>38 (46)</td>
<td>6 (50)</td>
<td>22 (39)</td>
<td>9 (64)</td>
<td>1 (100)</td>
</tr>
<tr>
<td>Female</td>
<td>45 (54)</td>
<td>6 (50)</td>
<td>34 (61)</td>
<td>5 (36)</td>
<td>0</td>
</tr>
<tr>
<td><strong>Haemoglobin genotype</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>SS/SS&lt;sup&gt;a&lt;/sup&gt;</td>
<td>71 (86)</td>
<td>11 (92)</td>
<td>48 (86)</td>
<td>12 (86)</td>
<td>0</td>
</tr>
<tr>
<td>SC</td>
<td>8 (10)</td>
<td>0</td>
<td>5 (9)</td>
<td>2 (14)</td>
<td>1 (100)</td>
</tr>
<tr>
<td>SB&lt;sup&gt;b&lt;/sup&gt;</td>
<td>4 (5)</td>
<td>1 (8)</td>
<td>3 (5)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td><strong>Hydroxyurea treatment at admission</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>38 (46)</td>
<td>4 (33)</td>
<td>28 (50)</td>
<td>6 (43)</td>
<td>0</td>
</tr>
<tr>
<td><strong>Hydroxyurea dose (mg/kg/day)</strong></td>
<td>17.9 (8-8-30.2)</td>
<td>18.8 (18-6-23.3)</td>
<td>18.2 (11-8-30.2)</td>
<td>13.7 (8-8-15-5)</td>
<td>--</td>
</tr>
<tr>
<td><strong>Weight (kg)</strong></td>
<td>68 (5-110)</td>
<td>32 (5-49)</td>
<td>71.8 (41-110)</td>
<td>71.5 (59-95)</td>
<td>85</td>
</tr>
<tr>
<td><strong>Vaso-occlusive crisis</strong></td>
<td>44/81&lt;sup&gt;c&lt;/sup&gt; (54)</td>
<td>6 (50)</td>
<td>34 (61)</td>
<td>4/12&lt;sup&gt;c&lt;/sup&gt; (33)</td>
<td>0</td>
</tr>
<tr>
<td><strong>Acute chest syndrome</strong></td>
<td>23/82&lt;sup&gt;c&lt;/sup&gt; (28)</td>
<td>2 (17)</td>
<td>17 (30)</td>
<td>4/13&lt;sup&gt;c&lt;/sup&gt; (31)</td>
<td>0</td>
</tr>
<tr>
<td><strong>Transfusion</strong></td>
<td>31 (37)</td>
<td>4 (33)</td>
<td>18 (32)</td>
<td>8 (57)</td>
<td>1 (100)</td>
</tr>
<tr>
<td><strong>Length of hospital stay (days)</strong></td>
<td>8 (2-37)</td>
<td>4 (2-10)</td>
<td>7 (2-35)</td>
<td>10 (4-37)</td>
<td>22</td>
</tr>
<tr>
<td><strong>Mechanical ventilation in the intensive care unit</strong></td>
<td>9/17 (53)</td>
<td>0</td>
<td>3/7 (43)</td>
<td>5/7 (71)</td>
<td>1 (100)</td>
</tr>
</tbody>
</table>

Data are n (%), n/N (%), or median (range). Percentages do not always equal 100% because of rounding. Ethnicity data were not collected in line with usual practice in France.

<sup>a</sup>Data for vaso-occlusive crisis were not available for two patients and acute chest syndrome not available for one patient.

<sup>b</sup>Simple transfusion or exchange transfusion (manual or automated) during the hospital stay.

<sup>c</sup>Hospitalisation was completed for 80 (96%) of 83 patients and is ongoing at the date of the notification for the other three.

<sup>d</sup>17 patients were admitted to the intensive care unit.
## COVID-19 and SCD – French Experience

<table>
<thead>
<tr>
<th>Age range (years)</th>
<th>Inpatients with sickle cell disease (n=83)</th>
<th></th>
<th>Hospitalised French population (n=17 745)*</th>
<th></th>
<th>p value†</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>ICU admission</td>
<td>Deaths</td>
<td>ICU admission*</td>
<td>Deaths†</td>
<td></td>
</tr>
<tr>
<td>All patients</td>
<td>17 (20)</td>
<td>2 (2)</td>
<td>6075 (34)</td>
<td>2891/42 212 (7)</td>
<td>..</td>
</tr>
<tr>
<td>0–14</td>
<td>2/12 (17)</td>
<td>0</td>
<td>32/110 (29)</td>
<td>1/592 (&lt;1)</td>
<td>0.72</td>
</tr>
<tr>
<td>15–44</td>
<td>7/56 (13)</td>
<td>0</td>
<td>514/2112 (24)</td>
<td>105/7524 (1)</td>
<td>0.039</td>
</tr>
<tr>
<td>45–64</td>
<td>7/14 (50)</td>
<td>2/14 (14)</td>
<td>3049/8422 (36)</td>
<td>1016/19 689 (5)</td>
<td>0.28</td>
</tr>
<tr>
<td>65–74</td>
<td>1/1 (100)</td>
<td>0</td>
<td>2480/7101 (35)</td>
<td>1769/14 405 (12)</td>
<td>..</td>
</tr>
</tbody>
</table>

Data are n (%) or n/N (%). *French general population younger than 75 years hospitalised with confirmed COVID-19 during the peak of the pandemic (April 7, 2020). †Comparison of ICU admission prevalence by age range between inpatients with sickle cell disease and the French general population hospitalised with confirmed COVID-19 (Fisher's exact test). ‡Death prevalence by age range among all confirmed inpatients with COVID-19 younger than 75 years from March 1, 2020, to April 14, 2020, in France.

**Table 2**: ICU admission in patients with sickle cell disease and COVID-19
Global Burden of Sickle Cell Disease During the COVID-19

COVID-19 and SCD: Registries and Surveys

Organizers
Dr. Julie Panepinto, Pediatric Hematologists
Dr. Amanda Brandow, Pediatric Hematologists
Dr. Ashima Singh, Epidemiologist and,
Dr. Lana Mucalo (Postdoctoral Fellow)
The Medical College of Wisconsin.
Global Burden of Sickle Cell Disease During the COVID-19

COVID-19 and SCD: Registries and Surveys

**Purpose:**
The goal of the registry is to report on outcomes of cases of COVID-19 in this population of patients.
Global Burden of Sickle Cell Disease During the COVID-19

COVID-19 and SCD: SECURE-SCD

Case Locations - USA
Global Burden of Sickle Cell Disease During the COVID-19

COVID-19 and SCD: SECURE-SCD
Global Burden of Sickle Cell Disease During the COVID-19

COVID-19 and SCD: SECURE-SCD

Age Distribution

Race & Ethnicity Distribution

Ethnicity: Hispanic/Latino, Not Hispanic/Latino, Not Reported, Unknown/Chose not to Answer

Number of Cases

Race Category: Black/African American, Other, Asian, White
Global Burden of Sickle Cell Disease During the COVID-19

COVID-19 and SCD: SECURE-SCD

Sickle Cell Disease Type (% of Total)

- Hgb SS disease: 70.98%
- Hgb SC disease: 20.09%
- Hgb S beta + thalassemia: 4.46%
- Hgb S beta zero thalassemia: 4.46%

Sex

- Female: 55.56%
- Male: 43.11%
- No Answer: 0.89%
- Other: 0.89%
Global Burden of Sickle Cell Disease During the COVID-19

COVID-19 and SCD: SECURE-SCD

COVID-19 Interventions

<table>
<thead>
<tr>
<th>Intervention</th>
<th>Yes</th>
<th>No</th>
<th>Not Reported</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventilation</td>
<td>59.56%</td>
<td>37.33%</td>
<td>3.11%</td>
</tr>
<tr>
<td>Transfusion</td>
<td>54.22%</td>
<td>27.56%</td>
<td>18.22%</td>
</tr>
<tr>
<td>ICU Admit</td>
<td>66.67%</td>
<td>11.11%</td>
<td>22.22%</td>
</tr>
<tr>
<td>Hospitalization</td>
<td>84.44%</td>
<td>15.56%</td>
<td></td>
</tr>
<tr>
<td>ED Visit</td>
<td>84.44%</td>
<td>15.56%</td>
<td></td>
</tr>
</tbody>
</table>

Average Hospital LOS: 8.17

Transfusion Type

- Exchange Transfusion: 27.38%
- Simple Transfusion: 72.62%
Global Burden of Sickle Cell Disease During the COVID-19

COVID-19 and SCD: SECURF-SCD

Disease Severity

- Mild: 52.89%
- Moderate: 18.67%
- Severe: 7.56%
- Asymptomatic: 16.00%
- Critical: 4.44%
- Unknown: 0%

Patient Death

- No: 91.56%
- Yes: 6.67%
- Unknown: 1.87%

Symptom Resolution

- Yes: 70.22%
- No: 14.67%
- Asymptomatic: 4.89%
- Unknown: 10.22%
Global Burden of Sickle Cell Disease During the COVID-19

COVID-19 and SCD: SECURE-SCD

Number of Patient Deaths

15
Global Burden of Sickle Cell Disease During the COVID-19

COVID-19 and SCD: Registries and Surveys

THANK YOU
Discussion

Please use the chat button in the toolbar on your screen to submit questions.
Update On The Global Coalition On SCD

Juliana Richardson
Senior Policy Advisor, US Department of Health and Human Services
The Coalition will harness the comparative advantage of each partner in a collective, ambitious but achievable goal of significantly reducing the morbidity and mortality of SCD in Africa.

Coalition initiatives will be implemented through, and inexorably linked with, the expansion of primary health care as a mechanism to achieve universal health coverage (UHC).

The initiatives will be centered on evidence-based practices that are highly cost-effective, feasible, and tailored to the specific needs and resources of individual countries.
Evidence-Based Interventions:

1. Define the Burden and Geographical distribution through Newborn or Early Infant Screening
2. Routine Preventative Care
3. Penicillin Prophylaxis through age five years
4. Affordable and Accessible Hydroxyurea
5. Pain Management
6. Functional Medical Record to enable a system of care
# Coalition Sub-Committees

<table>
<thead>
<tr>
<th>Treatment &amp; Protocol</th>
<th>Laboratory &amp; Diagnostics</th>
<th>Education &amp; Training</th>
<th>Implementation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Data Collection &amp; Disease Burden</td>
<td>Research</td>
<td>Donor/Private Sector Engagement &amp; Finance</td>
<td>Political Advocacy</td>
</tr>
</tbody>
</table>
OFFICE OF THE ASSISTANT SECRETARY FOR HEALTH

BRETT P. GIROIR, M.D.
ADM, U.S. Public Health Service
Assistant Secretary for Health

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Closing Remarks

Dr. Julie Makani
Associate Professor and Head of the Department of Hematology and Blood Transfusion at Muhimbili University of Health and Allied Sciences

Dr. Alexis Thompson
Section Chief of Hematology in the Department of Pediatrics Northwestern University School of Medicine