• No schedule of SOP revisions in 0/6 laboratories versus 4/6 laboratories: This clearly demonstrates an understanding that SOP’s need to be updated via a document management system.
• Scientist certified competences - 22 versus 22.
• Self-assessed improvement targets - 18 versus 15.
• Laboratory lead for QA - 3/6 versus 6/6: The importance of QA leadership for each laboratory identified prior to interim assessment.

Summary and Recommendations: Early evidence suggests that ARISE seedments have facilitated improved approaches to laboratory governance procedures and QA. Mentoring by EU partner laboratories will further enhance this. ARISE related workshops and train the trainer programmes will support capacity building and skills transfer. A final assessment will be undertaken at the end of the ARISE initiative.

Conclusion: The ARISE initiative will build on the early success of improvement in laboratory systems, to further support service development and capacity building. The target for each partner laboratory will be a timeline of application to full accreditation.

References
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S127 CONSORTIUM ON NEWBORN SCREENING IN AFRICA FOR SICKLE CELL DISEASE

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Background: Sickle cell disease (SCD) is an inherited blood disorder that causes abnormalities in the oxygen-carrying protein hemoglobin found in red blood cells, leading to severe long-term health effects. According to the World Health Organization, more than 300,000 babies are born each year with SCD globally with majority in sub-Saharan Africa failing to reach the age of five. With early identification, low-cost treatments known to be effective in high-income settings for several years, can improve the health of children with SCD. Yet, SCD has not received much funding in many African countries as a health priority limiting available diagnostic and treatment services for it.

The Consortium on Newborn Screening in Africa for SCD (CONSA), established in 2018, is a part of the American Society of Hematology’s broad initiative to strengthen the SCD response globally through advocacy and research generation.

CONSA Program Goals and Objectives
• Demonstrate the effectiveness of early identification and clinical interventions for newborns with SCD
• Create sustainable, expanded networks for newborn screening and clinical interventions
• Foster collaboration between African hematologists and public health services to develop an organized network of researchers for conducting quality studies and publishing results
• Increase hematology capacity throughout sub-Saharan Africa

CONSA introduces standard-of-care practices for screening and early intervention therapies (including antibiotics and malaria prophylaxis, folic acid supplements, family education and counseling, and immunizations) at participating clinical institutions, screening 10,000 – 16,000 babies per year in each country, and providing clinical follow-up for babies diagnosed with SCD. A shared registry captures data from CONSA institution sites, which will be used to estimate the prevalence of SCD in member countries and evaluate the effectiveness of the interventions for newborns to five years of age. Currently, CONSA is working in seven countries, Ghana, Kenya, Liberia, Nigeria, Tanzania, Uganda, and Zambia. Hematologists and public health officials participating in the consortium have mobilized networks of screening laboratories, SCD or pediatric hematology clinics, teaching hospitals, regional referral hospitals, universities, and satellite clinics to screen newborns and provide clinical services protocol. Alongside the research showcasing the health outcomes of newborns screened and delivered early interventions, the consortium is working to ensure the long-term sustainability of programs through government, corporate, and other partner support.

All country sites launched screening in 2021. As of November 15, over 17,000 babies have been screened with over 150 found to be living with SCD. Despite challenges from the COVID-19 pandemic, including population concerns of going to health clinics, need to protect SCD patients from transmission, and supply chain breakdowns, CONSA looks forward to continuing to expand newborn screening efforts for the next several years.

Conclusion: The presentation will provide an overview of CONSA’s goals and current work to screen and provide care for newborns with SCD, despite challenges from the COVID-19 pandemic. The presentation will also include details from the Nigeria clinical sites, case studies of current babies with SCD, and recent work done to strengthen Nigeria’s national newborn screening and clinical efforts.

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S128 CONTINUOUS QUALITY IMPROVEMENT IN PAEDIATRIC SICKLE CELL DISEASE

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Background: Sickle cell disease (SCD) is characterised by severe episodic pain requiring prompt and effective analgesia. However, SCD patients frequently experience poor quality of care due to lack of awareness of the condition among non-specialist staff, pre-conceived biases and unfounded allegation of drug-seeking behaviour. Several reports and surveys using Patient Reported Experience Measure (PREM) tools indicate widespread prevalence of ineffective pain relief in emergency department (ED), poor access to psychological therapies and poor funding for service development.

Aims: To use a validated PREM tool to survey patients or carers of sickle cell disease within the paediatric service of a Specialist Haemoglobinopathy Team. To use an established Continuous Quality Improvement (CQI) methodology (5-D’s—define, describe, design, deliver, digest) to ensure sustained improvement in patient-reported areas of service deficit.

Methods: The PREM survey was conducted as part of a network-wide initiative in 2018. The survey responses were analysed and problem scores created for specialist care, emergency care, ward-based care, information and support. These problem areas were categorised into domains where further improvement action was needed. CQI tools were used to map the patient journey within Emergency Department (ED) when presenting with pain followed by thematic analysis to identify potential improvements in the patient journey.

Results: The PREM survey analysis identified three domains for improvement:
• Domain A: To improve the effectiveness of pain relief and quality of empathic and informed care to paediatric sickle cell patients and their families in ED.

We developed multi-disciplinary teaching videos with patient involvement and patient information material based on ED/pain/sickle cell staff feedback. Excellent engagement from all staff groups obtained. Patient involvement in ED teaching video was well received. Thematic analysis of ED process mapping highlighted need for sustained staff education and re-writing ED pain protocol to align with current practice. Pre-conceived biases were identified among staff with respect to attributing drug-seeking behaviour to patients during severe sickle cell pain.
• Domain B: To improve the information presented to children and their families regarding SCD and the care services available in our trust

We developed child-friendly Information leaflets targeting primary school age and secondary school age readers.
• Domain C: To improve the availability of, and access to, mental health services for paediatric SCD patients.
We developed virtual parent support groups and psychological health screening for adolescent patients in clinics. We organised a “Tree of Life” (ToL) patient empowerment workshop. The parent support groups were well attended. Plans are in place to continue ToL workshops and support groups on a regular basis.

Future work: A rapid improvement workshop is being planned to engage a wider multi-disciplinary team to engender a culture of empathy in line with our trust values. Further input from workshop is expected to identify and implement sustainable goals. We plan to undertake a repeat PPR study using the end of this CQI project to understand the impact and the implemented improvements.

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S129 IMPLEMENTATION OF HYDROXYUREA THERAPY FOR SICKLE CELL DISEASE ON A LARGE SCALE IN GHANA
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Background: Hydroxyurea (hydroxycarbamide), has had a most profound and broad ameliorating impact on the clinical course of sickle cell disease (SCD). Although relatively inexpensive, hydroxyurea (HU) has not been widely available to the large majority of people with SCD who happen to live in low-income countries.

In 2018, in preparation for the establishment in 2019 of a broad-based Public Private Partnership (PPP) in SCD involving the Ghana government and Novartis, a group of parents Novartis to provide HU at a lower price for use in Ghana. Novartis produced 500mg capsule of HU and submitted it to Ghana Food and Drugs Authority registered the medicine for the specific indication of SCD in October 2018. The Sickle Cell Foundation of Ghana (SCFG), a partner in the PPP was tasked to develop the Ghana-Novartis Hydroxyurea-for-SCD Program (“Ahodwo [pr. A-ho-jol] Program”, meaning, “Program for Relief”).

The program was conceived as an implementation study to determine whether treatment with HU, specifically registered in Ghana for SCD, can be safely implemented and monitored on a large scale through an organized treatment program within the public health service in Ghana.

Methods:
1. Treatment Protocol: A team of Ghanaian SCD experts developed an HU-for-SCD dose-escalating, maximum tolerated dose (MTD) Ahodwo Protocol adapted for Ghana. A unique feature of the protocol is the selection of Hb level of 10g/dL as the primary goal of HU therapy with of a Therapeutic Dose (TD) defined as, “the dose at which Hb 10g/dL or higher is achieved and maintained over a period of 12 weeks”.

2. Treatment Teams: Established SCD Treatment Centres (SCD TC) were surveyed for patient numbers, age groups, Hb Phenotypes, and available laboratory services. Doctor-nurse-pharmacist teams were recruited from 11 TC located in four Regions of Ghana in Phase 1 of the Program for training. A year later, 9 smaller SCD TC were added to the program, in Phase 2, extending it to two additional Regions.

3. Ahodwo Program App: In order to register and guide healthcare professionals (HCP) on the protocol, register all subjects, assist with dosing calculations, and monitor the entire program, a secure, smartphone-based mobile application, Ahodwo Program App, was developed, tested, and deployed to all HCP in the program. Recording toxicity and reporting all expected/unexpected adverse events were mandated and reportable through the App.

4. Steering Committee (ST): A ST comprising clinician leaders of the TC was established; the ST held bi-weekly online review meetings for the first year and monthly thereafter. All HCP teams met every quarter.

Results: Table, below, lists the number and characteristics of subjects registered in the Ahodwo Program.

| Data Elements | Phase 1 | Phase 2 | TOTAL |
|---------------|---------|---------|-------|
| Number of SCD Treatment Centers (TC) | 11 | 9 | 20 |
| Average, No. of Subjects at TC | 3,357 | 291 | 3,648 |
| Median, No. of Subjects at TC | 305 | 32 | 182 |
| Range, No. of Subjects at TC | 256 | 26 | 88 |
| Female, No. (%) | 1,728 (47.4%) | 1,920 (52.6%) |
| Male, No. (%) | 1,561 (42.8%) | 792 (21.7%) |
| Age < 10yr, No. (%) | 1,295 (35.5%) |
| Age > 18yr, No. (%) | 792 (21.7%) |
| Presumed SCD-SS or S/beta-zero, No. (%) | 3,526 (96.7%) |
| SCD-SG, No. (%) | 122 (3.3%) |

On June 19th, 2021, World Sickle Cell Day, the government of Ghana announced the provision of HU for people with SCD through the National Health Insurance Scheme. Following preparatory meetings to establish the required regulatory standards, implementation of the national program is expected.

Conclusion: Our experience supports the tenet that hydroxyurea can be safely and effectively administered at population scale in a low-income country. Long-term sustainability in this setting is likely to be dependent on a government-supported access programme. The pioneering efforts of the government of Ghana to provide HU to its citizens with SCD are laudable and serve as a model to guide similar efforts in other low-income countries.

S130 PROCESSING SPEED DECLINES OVER TIME IN 4--25-YEAR-OLDS WITH SICKLE CELL DISEASE
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Background: Alongside physiological symptoms, young people with sickle cell disease (SCD) may also experience cognitive difficulties, including poorer processing speed. Processing speed develops rapidly from birth to around mid-childhood, with steady improvements thereafter into a person’s mid-twenties (Anderson, 2002). Nonetheless, little is known about the dynamic developmental trajectory of processing speed for young people with SCD.

Aims: This study, we aimed to investigate if the change in processing speed index (PSI) over time is significantly different between younger participants (aged under 8.99 years at first assessment) and older participants (over 9 years at first assessment) with SCD.

Methods: One hundred and five participants with SCD aged 4 -- 18 (N < 8.99 = 47; N > 9 = 58) at recruitment consented to follow-up IQ assessments (WPPSI-R, WISC-III, WASI-R or WASI-III) and MRI scans.