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PROCEEDINGS OF THE 2017 SICKLE CELL SUPPORT SOCIETY OF NIGERIA (SCSSN) BIENNIAL CONFERENCE – REDUCING THE BURDEN OF SICKLE CELL DISEASE (SCD) IN OUR COMMUNITIES

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INTRODUCTION

The Sickle Cell Support Society of Nigeria SCSSN is an umbrella body that brings together Nigerian doctors, scientists, NGOs and patients with interest in sickle cell disease (SCD), both within the country and in the Diaspora. The Society has made progress in advocacy for the control of (SCD), building collaborations for scientific research and establishing guidelines for uniform management of patients with the disease at primary secondary and tertiary health care levels. This has been done by working closely with the government and professional groups both within and outside the country.

The theme of the 2017 biennial conference was “Reducing the Burden of Sickle Cell Disease in Our Communities” with 4 sub themes: Multisectoral Approach in the Control of SCD, Early Detection of and Appropriate Intervention for SCD, New Treatments in SCD and Setting the Research Agenda for SCD in Nigeria. The Conference held at the Centre for Management and Environment Control, University of Nigeria, Enugu Campus, from the 23rd to 25th of August 2017. We hereby present the Conference proceedings

First Plenary Session: Multisectoral Approach in the Control of Sickle Cell Disease

The key paper was delivered by Dr Alayo Sopekan, the Desk Officer for Sickle Cell Disease, Non-Communicable Diseases Unit,

Federal Ministry of Health (FMOH). The session was chaired by Professor Godswill Obike. His paper was divided into three parts: a definition of multisectoral approach, its advantages and the role of FMOH as a major stakeholder in the control of SCD.

Multi-sectoral collaboration was described as a deliberate engagement among stakeholder groups or sectors to participate in dialogue, decision making and implementation of solutions in order to jointly achieve a policy outcome. In the case of SCD, it is to reduce its burden in Nigeria.

Multi-sectoral engagement leverages on the combined and varied strength in form of knowledge and expertise, reach and other resources to derive the maximum benefits. Effective multi-sectoral collaboration eliminates policy implementation barriers, facilitates scale-up and increases the impact that one sector or partner might have had alone. A successful multi-sectoral collaboration requires buy-in and commitment from all the stakeholders with coordination by the key sector. There must be an effective coordination mechanism in place such that decisions are applied and implemented across board. The key sector or partner must be able to identify opportunities for collaboration, mobilized the relevant sectors and set the agenda - as well as manage relationships. In addition to this, there must be a system to routinely monitor and assess the core mission deliverables and achievements.

SCD is a public health problem which is influenced by inter-related social, environmental and economic factors that can best be addressed with holistic, participatory multi-sectoral approach, which has been identified as one of the over-arching principles in the prevention and control of SCD.

The Federal Ministry of Health is a major stakeholder in the control of SCD in Nigeria. It provides leadership in the establishment and strengthening of a national multisectoral coordinated mechanism for prevention and control of SCD. It develops and implements a national multi-sectoral evidence base framework and action plan for prevention and control of SCD. It sets out priorities with time line and performance indicators. The FMOH has developed an evidence base National Guideline for management and control of SCD with the input of technical experts some of which are members of the SCSSN. There should be knowledge and information sharing to inform policy guidance and cost effective interventions. Finally, there must be closing of ranks among various SCD associations to ensure constructive and resourceful engagement with government.

Second Plenary Session: Early Detection and Appropriate Intervention for Sickle Cell Disease

The second Plenary Session was on early detection and appropriate intervention for sickle cell disease. The one and half hours session was chaired by Professor Angela Okolo and supported by Dr Titi Adeyemo. The main agenda during the session was robustly discussed by Professor Kwaku Ohene-Frempong of Ghana SCD Association. He talked on the need for Nigerian Network to set up new as well as monitor and regularly evaluate the existing Newborn Screening (NBS) Program for SCD in the country. He defined newborn screening as a public health program for early identification of conditions for which early and timely interventions can lead to the elimination or reduction of associated mortality, morbidity and disabilities. Citing the Ghana experience in about 21 years (1995 –

2016), out of a total of 481,302 babies who were screened, 8,252 (1.72%) had SCD, comprising of 4,259 with FS, 3,910 with FSC and 83 with FSA.

Three major threats to successful implementation of newborn screening were recognised. These were unstable Government budget support, absolute or near absolute reliance on foreign support for newborn screening in Africa and inability of the National Health Insurance Authority (NHIA) to meet their constitutional obligations. He proposed that efforts should be made to convert NHIA support to direct billing and that families should be encouraged to pay for newborn screening.

Report of the NBS activities at the Federal Medical Center, Keffi, North Central Nigeria was given by Dr Chinatu N Ohiaeri, the Centre Coordinator. The Sickle Cell Disease Care Centre in Federal Medical Centre, Keffi commenced operation in the year 2011 as one of the first four (4) Special Sickle Cell Disease Care Centres designated by the Federal Ministry of Health in 2011 to cater for North Central zone of Nigeria. The centre was specifically created for SCD screening for both neonates and older infants, as well as routine acute care and counselling. Other functions included organising training program for staff on the use of HPLC machine, institution of protocols for sample collection, processing and result analysis and interpretation as well as disclosure of results. From January to July, 2017, a total of 84 NBS had been done, with 17 (20.24%) being FAS (sickle cell trait) and none being FS (sickle cell disease). Overall, between 2013 and 2017, a total of 1203 samples were screened. One baby (0.24%) was identified as HbSS, 90 (21.69%) as HbAS, 2 (0.48%) as Hb S/* thalassaemia (SFA), 4 (0.96%) as HbAC and 3 (0.72%) as HbAD. With such low screening rates, it was obvious that the machine for NBS domiciled in the Centre was being grossly underutilized.

It was noted that NBS for SCD can reduce the burden of SCD in Nigeria, but presently, it appears there is no co-ordinated national NBS for SCD going on in Nigeria though Federal Government had produced a policy document

and guideline for SCD and NBS and had built and equipped six centres for dedicated NBS. The structures and facilities are decaying away in some of these centers. A way forward could be to charge a minimal fee as part of the ante natal fees from clients to ensure continuity of the program.

Third Plenary session: Setting the Research Agenda for Sickle Cell Disease in Nigeria and Opportunities for Capacity Building

The focus of the third and last plenary session of the Sickle Cell Support Society of Nigeria (SCSSN) was "Setting the Research Agenda for Sickle cell Disease in Nigeria and Opportunities for Capacity Building". Professor Jude Ohiaeri chaired the session with Dr Nkiru Odunukwe as co-chair. The discussion was led by the chairman of SCSSN, Prof. Adekunle Adekile, who opened with a brief mention of Nigeria's clear lead in the contribution to global sickle cell disease burden with an unmatched 91,011 out of the world's 305,772 annual SS births, its poor sickle cell disease survival rate of 10% compared 94% and 99% in the United States of America and United Kingdom respectively. He reminded participants of the objectives of SCSSN, which include advocacy, raising awareness about SCD, coordination of activities, capacity building and research. He stressed the need for national and international collaborations and encouraged participants to take advantage of various opportunities available to build capacity to help reduce the burden of with the Fiocruz Foundation, through which the government of Brazil gave scholarships to 8 Nigerians clinicians and scientists for MSc and PhD programs in the field of SCD in various Brazilian universities Prof. Adekile listed SCSSN research priorities as infections in SCD, hydroxyurea use, natural history studies for which Nigeria is well suited with its large patient numbers, ethno-pharmacology and last but certainly not the least, newborn screening for SCD.

Furthermore, he encouraged participation in workshops and conferences and listed some scholarship opportunities. Prof. Adekile enjoined professionals to choose an area of

sickle cell study to focus on, keep abreast of development in their area of interest and study the literature extensively. He advised researchers to find and work with a mentor, critically re-examine supposedly established truths and dogmas and increase the quality of their work and credibility by collaborating with carefully chosen teams. He concluded by revealing SCSSN's readiness to provide mentors, research and capacity building opportunities, and supervisors for postgraduate theses. He thereby unveiled the future agenda of the SCSSN in collaboration with the newly established Centre of Excellence for Sickle Cell Disease Research and Training (CESRTA) in Abuja, the West African and Global SCD networks.

The Chairman's call to focus and offer of collaborative opportunities was further buttressed by Prof. Ifeoma Okoye's presentation of events leading up to the establishment of the African Clinical Trial Consortium (ACTC), including plans to achieve the Clinical Trial Africa Vision 2020 birthed in 2016. This has been considered necessary to improve health care indices in Africa and reverse health tourism, thereby giving room for Africa's participation in global economy. She made a case for Africa's need for a clinical trial consortium, stating that the global clinical trial activity map featured only one African country which was the South African region where sickle cell is minimal in comparison to the burden in Nigeria. She shared the consortium's objectives and demonstrated clearly that SCSSN shared the vision and passion to increase capacity and build sustainable research networks for health innovation in Africa. She expressed ACTC's desire to collaborate with SCSSN and enumerated ways in which ACTC resources could be deployed to benefit the cause of SCSSN. She listed numerous benefits in the event of Africa's participation in the global clinical trial scene. These benefits were listed as improvement of public health, subsidy on healthcare, access to novel treatments, reversal of health tourism, improvement of the education sector, reduction of unemployment, reversal of brain drain, foreign revenue to boost

the economy and subsequently improvement of national reputation.

On whether Africa could deliver, Prof. Okoye noted the impediments but remained optimistic that they were not insurmountable. Some measure that would enhance Africa's performance were listed as establishment of regional centres of excellence equipped for clinical trials according to international best practice, increase commitment to uphold and enforce international property rights protection to ensure data protection, streamline and synchronise national regulatory activities with global guidelines. Prof. Okoye concluded by enjoining researchers to position themselves for Africa's preparation by obtaining Good Clinical Practice training and certification.

Dr Samuel Adegoke, thereafter presented his recently concluded experience with a SCSSN facilitated PhD opportunity at the Brazilian Health Ministry's FIOCRUZ Foundation in Rio de Janeiro. This opportunity was made possible by a memorandum of understanding (MoU) between SCSSN and FIOCRUZ which paved the way for Nigerian doctors and scientists to receive support and funding for MSc and PhD to study SCD in various Brazilian universities. These slots were however not filled to capacity as two candidates did not report for their programme. He described some of the skills acquired by the candidates, including transcranial Doppler (TCD) screening for stroke risk as very useful for Nigeria's needs and hoped that SCSSN would keep this bilateral relationship.

Dr. Ijeoma Diaku-Akinwumi thereafter discussed the importance and need for non-imaging transcranial Doppler (TCD) screening of children with sickle cell disease for ischaemic stroke risk. After a brief history of radiological screening for stroke risk, the anatomy and basis of stroke risk assessment by non-imaging TCD, the STOP guidelines for categorisation of stroke risk and interventions were discussed. Hydroxyurea's emergence and acceptability as an alternative to a less sustainable, less practicable chronic blood transfusion therapy to prevent stroke in children with high stroke risk were welcomed. Dr. Diaku-Akinwumi shared

the current state-of-the-art practice and work on stroke risk screening, showing how this low level of screening activity was grossly inadequate to cover the country's large population of children living with SCD. TCD screening is especially important given documented stroke incidence of 4-8%, mortality rate of up to 20% and recurrence rate in up to 75% of cases which leaves a quarter of survivors handicapped. She described the TCD technique, eligibility and interpretation, stating that the technique is easy to learn and the equipment simple, portable and easy to maintain. Participants were encouraged to seek to acquire equipment and expertise in TCD screening for stroke risk in order to attain standard practice in the management of SCD.

Dr. Titi Adeyemo discussed Nigeria's involvement in molecular and genomic SCD research with participation by Lagos University Teaching Hospital in Idi-araba, Lagos, University of Nigeria, Enugu and University College Hospital Ibadan. She opened the discussion by emphasising the need for empirical research to inform evidence-based interventions and policies relevant to African settings. She reminded participants of the factors known to affect the expression of SCD, with special focus on genetic modulators which could either be studied using genome wide associations (GWA) or by studying SNPs. She opined that an understanding of genomic modulation of SCD expression among Nigerians could change the face of SCD if defined and employed in effective interventions. Though a costly endeavour that African governments can ill afford on their own, Africa needs to embrace operational and genomic research as it needs the change it can bring and Africa has less mixed ethnicities presenting a unique opportunity for the study of SCD in its pure form. Dr Adeyemo acknowledged various barriers to Africa's participation and the dependence on external funding but proposed that strengthening of ethical, legal and logistic frameworks, improvement of political will, government involvement and private-public partnerships be employed to improve funding, foster a sense of ownership of the research findings and

enhance Africa's chances of sustaining research and health care improvement. She concluded by warning that research topics be carefully considered to ensure a contribution to disease management rather than academic curiosity, and proposed genomic basis of pain variability in SCD as an example of work that would contribute significantly to SCD management.

Prof. Obiageli Nnodu updated the conference participants on the infrastructure grant by the US National Institute of Health for the establishment of the Sickle Pan African Research Consortium with a hub in Tanzania and sites in Nigeria and Ghana. It is a four-year grant whose main objectives are to develop a SCD database, establish uniform multilevel standards of care, strengthen skills in health and research and to plan for future research. The consortium will work with existing SCD programs and ongoing activities in health, education and research. These objectives will be achieved by establishing a SCD database of 6000 paediatric and adult sickle cell patients in Nigeria within the institutional health information management systems. Locally-appropriate standards of care will be determined based on available resources. Working with existing programs, SPARCO will increase the quantity and quality of skilled healthcare professionals working in SCD and build on existing research activities to plan for clinical and implementation studies. In Nigeria, the SPARCO Co -Principal Investigator Professor Obiageli E. Nnodu, will work with members of Sickle Cell Support Society of Nigeria within the Clinical Centres of the Network to achieve the set objectives which will help to reduce the burden of SCD in Africa and increase the capacity for research that will contribute to scientific knowledge to find a cure for SCD.

She explained the SCSSN's unique position to strengthen Nigeria' position and contribution in the Consortium by elucidating SCSSN's research agendas, which broadly shares some common research themes by SPARCO. She elaborated on the various promising funding opportunities available to SCSSN for co-

ordinated SCD research and skills development with SPARCO. She also brought to participants' notice the essence of SCSSN research priorities which were captured by underlisted research questions agreed upon at a workshop sponsored by the British High Commission in Abuja in 2015 to establish research collaboration between the SCSSN and United Kingdom partners:

1. What sorts of therapies are feasible and reasonable for reducing organ damage in SCD sufferers?
2. How can we screen for SCD in an affordable way using existing public health system models in Nigeria/West Africa?
3. How can we reduce the number of sickle cell births?
4. How does sickle cell vary across Nigeria, and how can we document those variations?
5. What is the socio-economic cost of sickle cell in Nigeria?

She urged members to consider and champion these in order to generate pilot data which can become useful as the foundation for future research in SCD as being laid through SPARCO.

This robust session was concluded with videos and demonstration of 2 point-of-care test kits for diagnosis of SCD, SickleScan and HaemoType SC. These immunogenic test kits did not require power supply and proved easy to use and interpret in the community with minimal training. This raised hope for its use in newborn screening, encouraging conference participants to eagerly enlist in a co-ordinated nationwide validation of HaemoType SC in the field among infants 9 months of age and less in their communities.

The plenary session's rapporteurs Dr.Umar Kangiwa and Hezekiah Isa summarised and closed the conference deliberations thereafter and a communique of the conference resolutions was drafted.

CONCLUSION

The third Conference of Sickle Cell Support Society of Nigeria (SCSSN) was successfully held at the Enugu Campus of the University of Nigeria with the theme: "Reducing the Burden of Sickle Cell Disease in our Communities" from Wednesday, 23rd to Friday, 25th of August, 2017.

Three plenary sessions were coordinated by erudite scholars and renowned researchers in the field of SCD. Highlights of the plenary sessions included discussions on:

- Multi-sectorial approach in the control of SCD
- Early detection and appropriate intervention in the control of SCD
- Setting up a new-born screening program for SCD
- Setting the research agenda for SCD and opportunities for capacity building
- Reports from the Zones
- Genomics research on SCD and update on the NIH-funded Sickle Pan African Research Consortium.

The Sickle Cell Support Society of Nigeria noted that despite an enabling policy and purchase of equipment for new-born screening in the MDG, SCD Screening Centres, implementation has been fraught with challenges of procuring reagents resulting in poor utilisation and redundant machines. Availability of Transcranial Doppler machines for stroke risk assessment and primary stroke prevention is still poor. It was agreed that the most practical intervention for stroke prevention is hydroxyurea therapy since chronic blood transfusion seems herculean and impractical in most of our communities. It was noted that SCD-related activities of NGOs and researchers in the country are currently not co-ordinated.

The dearth of funding for research and lack of an enabling environment were discussed at length. More co-ordination in research and NGOs' activities is necessary to make impact. It was suggested that Nigeria and its government take responsibility for the sickle cell problem rather than rely on international funding while

missing components of comprehensive SCD care should be addressed.

RECOMMENDATIONS

The participants rose from the AGM with the following recommendations:

1. The Sickle Cell Support Society of Nigeria (SCSSN) noting the lack of data on the true prevalence of SCD in Nigeria is ready to partner with the Federal Ministry of Health to obtain accurate data for SCD in Nigeria
2. The SCSSN seeks an end to the current redundancy of the machines for new-born screening in the MDG Sickle Cell Centres and calls on the Federal Ministry of Health to make annual budgetary provision for reagents and consumables for new-born screening, as well as recalibration and preventive maintenance of the current machines at the centres.
3. The SCSSN is ready to collaborate with and coordinate various SCD-related NGOs to present a united front to tackle SCD in Nigeria and encourages collaboration among researchers. The SCSSN recommends that TCD machines and expertise be made universally available to offer standard-of-care for the prevention of stroke in children with SCD.
4. The SCSSN noted the lack of data on SCD and recommends a centralised sickle cell registry fed by the new-born screening programme and community screening in the different zones.
5. The SCSSN calls on the Government to increase research funding for sickle cell disease.
6. The SCSSN wants a co-ordinated judicious deployment of resources as recommended to make greater impact in reducing the burden of SCD in Nigeria.

The Chair of the 2017 SCSSN Conference was Professor Obiageli Nnodu, the Secretary, Dr Funke Lawson and the Chair, Local Organizing Committee was Professor Iheanyi Okpala.