What every physician should know about the national guidelines for the control and management of sickle cell disease and the parent handbook for sickle cell disease in Nigeria

Sickle cell disease (SCD) is a genetic disorder that is caused by the inheritance of HbS, which results from the substitution of valine for glutamic acid in the 6th amino acid position of the β-globin chain. Individuals who are homozygous for this substitution have sickle cell anemia (SCA) which is the most severe form of SCD.[1] The greatest burden of SCA is in sub-Saharan Africa (SSA), where approximately 75% of the global 300,000 births of affected children occur with 50–80% of the children dying before adulthood.[2,3] The World Health Organization (WHO) estimates that 70% of deaths from SCD in Africa are preventable with simple and cost-effective interventions.[3,4]

Since the first formal report of SCD more than a century ago,[4,5] and the era of confusion and uncertainty that characterized its discovery,[5] various landmark efforts have been made to unravel the complexity and pathophysiology of the disease. Laudable among such efforts include the elucidation of its genetics by Pauling *et al*. in 1949,[6] followed by the characterization of the involved mutation and its position in the polypeptide chain.[7] However, the enactment of the SCA Control Act by the US Congress in 1972 was a major stimulus for research and the acquisition of various management strategies.[5] This did not only lead to increased awareness about SCD but also increased research funding by various reputable bodies such as the USA National Institutes of Health, the British Medical Council, and the Wellcome Trust.[5] The outcomes of these efforts are the various guidelines available in most developed countries as well as some novel strategies and concepts for managing SCD. Penicillin prophylaxis in early childhood, comprehensive care, Transcranial Doppler screening for stroke risk, and use of hydroxyurea are good examples.[5,8] Although the implementation of these efforts has led to a drastic reduction in both the morbidity and mortality of SCD patients in the developed world,[9,10] the outlook in most developing countries remains gloomy. This has been attributed to the lack of sustainable efforts at addressing the huge burden of the disease through the use of simple and cost-effective measures in these resource-poor countries.[3,11-13]

Although Nigeria has the highest burden of SCD in the world, with about 2% prevalence in newborns,[14] there has been lack of coordinated national effort in combating the disease in the country until recently. Nothing drives home this point than the warning by Molineaux *et al*.[15] in 1979 on SCD in Nigeria, where they wrote among other things that "*There is no other known inherited disorder present at such high frequency in a large population and of comparable severity as sickle cell anemia in Africa. With rising standards of living and control of malaria, sickle cell anemia will become an immense medical, social, and economic problem throughout the continent*." Indeed and true to their prediction, SCD has not only become an immense medical problem[16-18] but also a huge economic burden in Nigeria.[19,20] Recent work by Piel *et al*. has also reinforced the predicted exponential increase in the prevalence of the disease in SSA, especially Nigeria, in the near future.[21,22]

Repeated efforts by the WHO[2,23] and the recognition of SCD as a disease of public health importance by the United Nations in 2008, which culminated in the designation of June 19, of each year as World Sickle Cell Day, have instigated more action on SCD in Nigeria. The formation of the Nigerian SCD network now known as sickle cell Support Society of Nigeria in 2010, brought together many individuals, experts, and other interested bodies to fight the SCD burden in Nigeria.[14,23] The federal government responded in the same year by assigning a desk officer solely for the control of SCD in the Noncommunicable Diseases Division of the Federal Ministry of Health. In 2012, six Federal Medical Centers, one each from the six geopolitical zones were empowered to run dedicated clinics, provide newborn screening and other programs for the management and control of the disease. The launch of the first national guidelines for the control and management of SCD and the parent handbook for SCD in Nigeria were the climax of recent efforts in the country in this regard.

The national guidelines for the control and management of SCD (NGCMSCD) is the first of its kind in Nigeria. The document was launched on November 28, 2014, by the federal government.[24] It is a product of well-researched contributions from eminent scholars, professional bodies, and reputable nongovernmental organizations in the field of SCD care based on current best practices in the world.[25] The NGCMSCD comprises major recommendations for the

care of persons living with SCD in Nigeria. The guidelines consist of nine chapters, each dealing with specific aspects of SCD. The first two chapters provide background information on SCD and its diagnosis, the third chapter summarizes its common key manifestations, while specific indications for some form of specialized treatments are provided in the fourth chapter. The fifth chapter focuses on the management in pregnancy and the 6th chapter highlights management of chronic complications of the disease. Guidelines on the approach to growth monitoring, nutrition, immunization, and other cares in steady state are provided in the 7th chapter. Finally, borrowing from the famous Harveian Oration which says "Prevention is better than Cure"[26] the last two chapters focus on preventive strategies.[25]

On the other hand, the parent handbook for SCD in Nigeria[27] is an adaptation of a similar handbook for parents of patients with SCD in the USA where it has been in use for more than two decades. The adapted version of the book is packaged into two volumes (Parts I and II). While Part I contains information for parents on the care of children aged up to 6 years, part II deals with SCD in older children. Specifically, the book focuses on information on common problems in children with the disease and provides appropriate answers on steps to be taken by parents in responding to these problems. In addition, it provides answers in simple language to some frequently asked questions by caregivers including dispelling some myths about the disease. Furthermore, the book provides adequate information to the parents in lay language on danger signs in children with SCD.

The launch of these documents is strategic and timely as they readily complement each other. While the NGCMSCD is mainly targeted at healthcare professionals by providing state-of-the-art management approaches for SCD patients, the handbook provides adequate information in lay language to the parents and other nonmedically inclined caregivers, thus providing holistic care to the patients by bridging the information transfer gap to parents and other caregivers. This is very important given that home care by parents or caregivers plays a vital role in the wellbeing of SCD patients. There is therefore the need for all stakeholders, especially physicians, to key into their use and deliver quality care to SCD patients in the country. It is hoped that this will lead to better utilization of available local resources to cater for individuals with the disease in the country. Hopefully, these efforts should culminate in reduced morbidity and mortality caused by SCD. It therefore behoves every healthcare practitioner involved in the care of SCD patients in Nigeria to avail themselves of the contents of these vital documents and help their patients get the best care available. In order to reach the targeted end users, there is the need to give the documents wide publicity through print and electronic media. In addition, appropriate government agencies should make copies freely available to various health institutions and professional bodies such as the Nigerian Medical Association, Paediatric Association of Nigeria, Nigerian Society for Haematology and Blood Transfusion, and the Medical and Dental Consultants' Association of Nigeria who are involved in the care of patients with SCD. Furthermore, government should make the parent handbooks more widely available by translating them into the various major local languages so that the lay public will be more familiar with their contents. Currently, the electronic copies of the handbook and the national guidelines can be accessed online at the following links www.scsn.com.org and www.schafng.org. Finally, we advocate that these vital documents should be constant companions of every Nigerian physician involved in SCD care for easy and ready consultation.

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Conflicts of interest

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O S Olatunya, A D Adekile[1]

Department of Paediatrics, Faculty of Clinical Sciences, College of Medicine, Ekiti State University, Ado-Ekiti, Ekiti State, Nigeria, [1]Department of Paediatrics, Faculty of Medicine, Kuwait University, Safat, Kuwait

Address for correspondence:
Dr. O S Olatunya, Department of Paediatrics, Faculty of Clinical Sciences, College of Medicine, Ekiti State University, Ado-Ekiti, Ekiti State, Nigeria.
E-mail: ladeletunya@yahoo.com

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