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## Health Professional Treatment Practices for Pediatric Sickle Cell Disease in Nigeria

Natalie Wichelt  
*University of Nebraska Medical Center*

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Running Header: Health Professional Treatment Practices for Pediatric Sickle Cell Disease in Nigeria

## Health Professional Treatment Practices for Pediatric Sickle Cell Disease in Nigeria

Natalie Wichelt  
MD/MPH Student

**Abstract**

Sickle cell disease (SCD) is caused by a genetic defect that results in abnormal hemoglobin genes that can cause devastating health effects like chronic hemolytic anemia and vaso-occlusive crises. Additionally, sickle cell disease can cause defects in the immune system, leaving those with the disease highly susceptible to a variety of different infections. In the pediatric population specifically, the complications of sickle cell disease contribute significantly to the under-five mortality rate of Nigeria. In response to the Millennium Development Goals 4, 5 and 6, as well as with growing national concern over the challenges faced by sickle cell disease there was a collaboration with the Federal Ministry of Health (FMOH) with the Millennium Development Goal office to create a national standard of care for sickle cell disease treatment. This document was created and published on November 28, 2014, and since then there has not been a study done to determine if the national guidelines that were created have been adopted or if there were barriers preventing this from occurring.

The purpose of this study was to seek out health providers in Nigeria from different parts of the country, with different clinic and provider demographics to examine what current practices are being used and how they compare with the guidelines. The study aimed to provide a better understanding of providers awareness of the national guidelines for treatment, what the proportion of their patients had treatment that followed the guidelines, if their prescription habits aligned with the guidelines and if not, what the barriers to following the national guidelines were.

To investigate how provider practices compared to the national guideline for SCD, a comprehensive online survey was distributed to 96 Nigerian healthcare providers at the joint 14<sup>th</sup>

Triennial Congress of Union of National African Paediatric Societies and Associations (UNAPSA) and the 49<sup>th</sup> Paediatric Association of Nigeria (PAN) Annual General Meeting and Scientific Conference in Abuja, Nigeria from January 24-26, 2018. Of the 96 participants that were contacted, 31 fully completed the survey with a response rate of 32.3%. Important to note was that the majority of the providers, 58.1%, were not aware that there was a national guideline for sickle cell disease treatment available. The results of the study show that while specific treatment practices like malarial prophylaxis and folic acid supplementation are widely used, there is still much room for improvement for newborn screening, pneumococcal vaccination and hydroxyurea. In addition to the elements of the national guidelines, there was also interest in the role of other medications like ciklavit and herbal preparations when it came to sickle cell disease for which there was little provider support. This study will provide the foundation for future interventions to increase the awareness and provider knowledge of the national guidelines for pediatric sickle cell disease treatment to provide better streamlined and universal care for those affected by this disease in Nigeria.

## **1. Introduction**

Sickle cell disease (SCD) is the result of a genetic mutation that is one of the leading causes of disease in the world, with some of the highest rates of prevalence being in Nigeria, where it remains a leading killer of children under five years old (Galadanci 2014). Sickle cell disease is categorized as a chronic hemolytic disorder. As a result of the mutation, there is a change in the shape of the red blood cells and they take on a deformed or sickle shape. This sickle shape causes many different complications in the health of patients leading to vaso-occlusive events, pain crisis and hemolysis (Galadanci 2014). According to Adewoyin in the

*Management of Sickle Cell Disease: A Review for Physician Education in Nigeria*, “Sickle cell disease affects about 2 to 3% of the Nigerian population of more than 160 million” (Adewoyin 2015). The majority of patients that have sickle cell disease are not aware of their status, until they experience a complication from the disease and end up in a general clinic or hospital (Adewoyin, 2015). This highlights the importance of understanding what the treatment habits are of not only those who are in specialized sickle cell disease clinics, but also those who are practicing in the ER, general clinics and hospital settings.

Given the medical complications that can occur with sickle cell disease patients who develop bacterial infections, malaria and HIV infection, the management and care of sickle cell disease can be very complicated. It is important to note that while the heterozygote form of the sickle cell trait can afford some protection against malaria, the homozygous states like that seen in sickle cell disease can make the effects of malarial infection more severe (Galadanci 2014). Some of the advised methods of management include ruling out malarial infection, folic acid supplements, monitoring and caring for anemic states and using antibiotics efficiently when appropriate. By understanding health care decision making, the disease burden of sickle cell disease in Nigeria can be improved through better care. An evaluation of healthcare practices surrounding sickle cell disease in Nigeria was done in a study by Galadanci; in which they surveyed various sickle cell specific institutions throughout the country to gather data on the protocol for patients with sickle cell, including diagnosis and treatment (Galadanci, 2014). They found that it was standard to give folic acid and malaria prophylaxis but that there was a gap in care for penicillin prophylaxis and pneumococcal vaccines are not routinely provided. Diagnostically, the study showed that none of the 11 institutions that participated had any established newborn screening program. The foundation of this study was an important step

towards establishing better sickle cell management through the 2010 Nigerian SCD Network who conducted this study to get data on current care and develop a better plan for future care.

In order to face the growing burden of pediatric sickle cell disease on the under-five mortality, Nigeria passed a national guideline for treatment practices in November of 2014 titled the National Guideline for the Control and Management of Sickle Cell Disease. This document provides a foundation for sickle cell screening, treatment and complication prevention (Nigeria 2014). However, no comparison of medical professional treatment of sickle cell disease has been done since the passage of this national guideline.

By having a standardized treatment plan, there could be better response and treatment of kids with sickle cell disease. The purpose of this study is to understand what current healthcare provider knowledge is of the national guidelines and how provider treatment practices compare to the standard of care described in the national guidelines. Additionally, there was a desire to understand if there was a gap between the standard and actual current healthcare practice to identify what the barriers to care were and why certain practices were not always being carried out. Since the guidelines were created, a study to see the impacts on current practice has not been done and the results will be important in determining next steps to creating better knowledge and standardized rules of practice to improve the health of pediatric sickle cell disease patients. This project will help to understand what current treatment plans are being used, the factors that are impacting these choices and ways that better care can be provided by understanding the current barriers. Additionally, this study extends beyond sickle cell specific clinics to include the full range of health care facilities available. By understanding these gaps in care and why they exist, we can have specific areas to focus on to create successful interventions to hopefully lead to

universal standards of care that are also lived out in actual practice to reduce the under-five mortality due to sickle cell disease.

## **2. Methods**

### **2.1 Study Design**

The design of the study was a questionnaire based on convenience sampling of health care professionals. The study was conducted in partnership with the International Foundation Against Infectious Disease in Nigeria (IFAIN). IFAIN has the mission of reducing and better understanding infectious disease burdens in Nigeria. IFAIN is a Nigerian non-profit, non-governmental organization that is an affiliate of UNMC. IFAIN was established in 2012 with support from UNMC to manage UNMC research projects in Nigeria.

### **2.2 Study Population**

Medical professionals in Nigeria that work in a pediatric setting were the participants targeted for this study. The study sample resulted from emails that were collected at the joint 14<sup>th</sup> Triennial Congress of Union of National African Paediatric Societies and Associations (UNAPSA) and the 49<sup>th</sup> Paediatric Association of Nigeria (PAN) Annual General Meeting and Scientific Conference in Abuja, Nigeria from January 24-26, 2018. Additionally, providers through medical clinics in Nigeria that have existing collaborations with IFAIN participated. Of the 96 total participants that were contacted, there was a 32.3% response rate with 31 fully completing the survey.

### **2.3 Data Collection Methods**

The survey was completed through the online REDcap system to keep the information secure and only available to those designated at IFAIN and UNMC. The survey had

a total of 58 questions that took around 15 minutes to complete, a copy of the detailed questions can be found in Appendix A. The question types included direct yes/no, multiple choice, multiple check boxes and fill in the blank responses. The survey was divided into the following sections; Clinic Characteristics, Provider Demographics, Guideline Knowledge, Provider Treatment Practices and Furthering Education. This was done in an attempt to understand the multiple factors that are involved in knowing about and practicing according to the recommendations set out in the national guidelines. Once the survey was completed by the participants, they were compensated for the data usage required to participate. Minutes were added back onto the device that was used to complete the survey by an IFAIN representative in Nigeria who received the phone numbers through the secure REDcap database

#### **2.4 Data Analysis**

Descriptive statistics were used to compare the treatment habits medical professionals in Nigeria to treatment prophylaxis recommended in the national guidelines. The analysis provided information regarding knowledge of national guidelines and provider prescription habits related to the prophylactic measures outlined in the national guidelines for pediatric sickle cell disease patients. The survey compared results between pediatricians, general practice physicians, pediatric residents, pediatric subspecialists that are not hematologists and medical officers. Nurses and community health workers were not participants of this study. The sites that providers worked in included the hospital, general clinic, ER and specialty clinics. The primary population demographics, public or private and rural or urban as institution types of the healthcare providers was also explored. For the questions regarding the proportion of patients on specific prophylactic treatments the following categories were used, 0-25%, 26-50%, 52-75% and 76-100%. There were some cases where there were extremes in data with either very few



patients on treatments or almost all of the patients on treatments and the categories were then divided up accordingly, i.e. <10% as a category or 90-100% as a category. The results of the survey can be used to identify gaps between actual healthcare practices and the national guidelines to aid in policy analysis and interventions to increase the efficiency and uniformity of treatment of sickle cell disease.

### **3. Results**

There were 96 surveys distributed with 32 surveys submitted and 31 fully completed survey responses with participants representing various different clinical and provider demographics with a response rate of 32.3%. The survey was divided into the five question sections that the results will follow accordingly; Section I: Clinic Characteristics, Section II: Provider Demographics, Section III: Guideline Knowledge, Section IV: Treatment Practices, Section V: Furthering Education.

#### **3.1 Section I: Clinic Characteristics**

In this section, the purpose was to gather information to describe the different types of facilities and clinic backgrounds that our providers were working in. This information could then be used to understand if there was a difference in practice that was related to the clinic characteristics. The results of the survey found that of the 31 participants, 20 (63.5%) primarily practiced in a teaching hospital, 3 (9.7%) worked in a hospital inpatient unit, 3 (9.7%) in hospital outpatient, 2(6.5%) in an emergency room setting, 1(3.2%) in a general clinic and 2 (6.5%) in other settings. The other settings that were described were a general hospital and a federal medical center.

When it came to the characteristics of the clinic, providers could choose more than one option; 2 worked in an adult facility, 13 in a pediatric facility and 17 in an adult and pediatric

clinic. There were 13 providers that marked that they worked in a public facility and 2 that worked in a private facility. The majority of providers worked in an institution that was urban, 28 (90.3%) while 3 (9.7%) were from a rural facility. All of the institutions kept medical records, and of those 17 (54.8%) of them kept a database of their sickle cell disease patients.

The diagnostic equipment for sickle cell disease varied depending on the clinic. Of the equipment available at the facility participants selected all the options that they had available at their facility, all had x-ray available, 50% had a transcranial doppler, 60% had a CT and 26.7% had an MRI machine. For the diagnosis of sickle cell disease in particular, the main method providers stated that of the diagnostic methods used 87.1% of their facilities used electrophoresis (cellulose acetate, citrate agar), 9.7% used capillary electrophoresis, 9.7% used HPLC and 3.2% not being sure. The majority, 23 (74.2%), of providers stated that the facilities did not have a genetic counselor available at the institution, with only 3 (9.7%) reporting they did and 5 (16.1%) not knowing. Lastly, given the emphasis of newborn screening in the national guidelines, we were curious to know if newborn screening was done at their facility and found that 27 (87.1%) do not conduct newborn screening, 3 (9.7%) do and 3 (3.2%) did not know.

### **3.2. Section II: Provider Demographics**

In this section, we wanted to understand the different types of providers that were completing the survey and gather information on their respective backgrounds to see if this influenced their knowledge of national guidelines for treatment and their treatment practices. Of those who completed the survey, 17 (54.8%) were male and 14 (45.2%) were female. There was a range of ages 31-55 represented with the mean age of providers being 44 with a standard deviation of 6.68. The range of practice years was 5-31 years with an average years of practice of 18.9 years

with a standard deviation of 7.53 . The education levels of the survey participants showed that 17 held an MBBS, 18 were postgraduate fellows, 3 an MD and 5 had a Master's degree, with some providers holding multiple degrees. There were graduates from various institutions including 7 from the National Postgraduate Medical College of Nigeria (NPMNC), 3 from West African College of Physicians, 17 from African Universities and 3 from International Universities. The providers were from different areas of Nigeria including Kano, Kwara, Oyo, FCT/Abuja and Lagos.

Professionally, 22 (71%) of the survey participants were identified as pediatricians, 3 (9.7%) as a pediatric resident/registrar, 3 (9.7%) as a pediatric subspecialist not hematologist and 3 (9.7%) as a medical officer. The participants saw range of pediatric sickle cell disease patients from 0-40 per week with an average 8.63 pediatric sickle cell patients per week and a standard deviation of 10.99.

### **3.3 Section III: Guideline Knowledge**

To assess the awareness and use of the national guidelines for sickle cell disease treatment, we asked the participants about the extent of their guideline knowledge. We found that 15 (48.4%) of the participants had heard about a Nigerian National Guideline for Sickle Cell Disease Treatment. However, when asked about the National Guideline for the Control and Management of Sickle Cell Disease (2014), 18 (58.1%) had not heard of the national guideline. When questioned about the frequency that participants followed the national guidelines for sickle cell disease treatment, the majority 14 (45.1%) did not know how often they followed the guidelines, 9 (29%) did often, 3 (9.7%) never did, 2 (6.5%) sometimes did, 1 (3.2%) always did, 1 (3.2%) rarely did and 1(3.2%) did not answer. In order to determine possible barriers to following the national guidelines in practice, participants were asked to check possible reasons

for not always following the guidelines. The most commonly identified barriers were asked in a check all that apply format with the following checked “I did not know that there were national guidelines” 11 (47.8%), “I do not have a copy of the guidelines” 8 (34.8%) and “I am not the primary SCD patient provider” 5 (21.7%). Also indicated as barriers, were preference for other guidelines 2 (8.75%), vaccines not being available 2 (8.7%), labs not being available 1 (4.3%) and not having received sickle cell disease patient care specific training 1 (4.3%).

### **3.4 Section IV: Treatment Practices**

The core of the practices that were focused on in the survey were the prophylactic measures recommended by the national guidelines including penicillin prophylaxis, malarial prophylaxis, pneumococcal vaccination, folic acid supplementation and hydroxyurea. In an attempt to gather information on other treatment practices, ciklavit and herbal preparations were also included in this survey. The most widely followed treatment practices were the use of malarial prophylaxis and folic acid supplementation. There was some use of penicillin prophylaxis and pneumococcal vaccination, a wide range of practices for hydroxyurea and little use of ciklavit or knowledge of herbal preparations.

Each subsection can be divided into the following for each treatment practice as the questions were asked in terms of patient population, frequency of provider prescriptions and a qualitative assessment of why a practice was not “always” done. For penicillin prophylaxis, 14 of the respondents estimated that <10% of their patient population was taking penicillin prophylaxis, followed by 9 that did know, 1 that estimated 11-25%, 1 that estimated 51-75%, 2 that estimated 76-100% and 1 that did not answer. When asked about their prescribing habits for penicillin prophylaxis, 7 (22.5%) never did, 6 (19.4%) sometimes, 6 (19.4%) rarely did, 5 (16.1%) often

did, 4 (12.9%) always did and 3 (9.7%) did not answer. When asked why they did not always prescribe penicillin prophylaxis for their pediatric sickle cell disease patients there were 17 responses, the majority stated that it was not a part of the routine practice/treatment protocol 4 (23.5%), there was not enough evidence for the success of this treatment practice 3 (17.6%), they referred out their SCD patients 3 (17.6%), the pneumococcal vaccine had already been given 1 (5.9%) or that they did not know that they needed to 1 (5.9%).

For malarial prophylaxis, 24 of respondents estimated that 76-100% of their pediatric sickle cell disease patients were taking malarial prophylaxis, 1 estimated that 11-25% of their patients were, 3 estimated 51-75% of their patients were, 1 did not know and 2 did not answer. When asked about their prescribing habits for malarial prophylaxis, 27 (87.1%) always did, 1 (3.2%) often, 1 (3.2%) sometimes did and 2 (6.5%) did not answer. For the pneumococcal vaccine, 14 did not know how much of their patient population had received the vaccine, 4 estimated <10%, 4 estimated 51-75%, 3 estimated 11-25%, 3 estimated 26-50% and 2 estimated 76-100%. When asked about their prescribing habits for the pneumococcal vaccine, 9 (29.0%) always did, 6 (19.3%) sometimes did, 5 (16.1%) often did, 3 (9.7%) never did, 3 (9.7%) rarely did, 2 (6.5%) said it was unavailable and 3 (9.7%) did not answer. Of those who did not answer that they always prescribed the pneumococcal vaccine for their pediatric sickle cell disease patients there were 15 responses, the majority 9 (60%) indicated cost as a major barrier, 3 (20%) stated that their patients were already immunized, 1 (6.7%) said that the vaccine was unavailable, 1 (6.7%) that they referred their SCD patients elsewhere and 1(6.7%) answered other.

For folic acid supplementation, 26 respondents estimated that 91-100% of their pediatric sickle cell disease patients were taking folic acid supplementation, 2 estimated 70-90%, 1 did not know and 2 did not answer. All of the providers, 100%, said that they always prescribe it. For

hydroxyurea, the majority of respondents (11) estimated that <10% of their patients were taking it, 10 did not know, 5 estimated 26-50%, 2 estimated 11-25%, 1 estimated 51-75%, 1 estimated 76-100% and 1 did not answer. When asked about their prescribing habits for hydroxyurea, 8 (25.8%) sometimes did, 6 (19.3%) often did, 6 (19.3%) rarely did, 4 (12.9%) never did, 4 (12.9%) found it unavailable and 3 (9.7%) did not answer. When asked why they did not always prescribe hydroxyurea for their patients there were 20 responses, the majority of respondents cited the prescription guidelines based on disease severity 11 (55.0%), referral to a specialist 4 (20%), cost as a barrier 3 (15%) and 2 (10%) stated other.

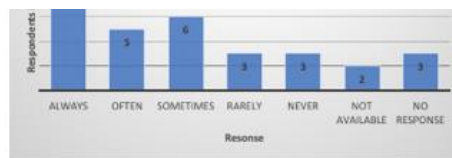
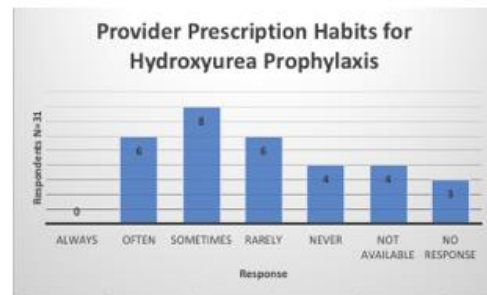
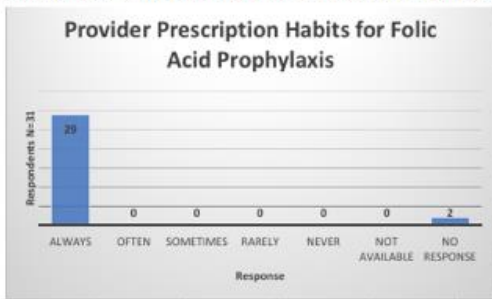
Figure 1: Graphs Depicting Provider Prescription Habits for SCD Complication Prophylaxis from Survey Sample of Nigerian Healthcare Providers, 2018 (Respondents N=31)

Graph 1: Provider Prescription Habits for Penicillin Prophylaxis

Graph 2: Provider Prescription Habits for Malarial Prophylaxis

Graph 4: Provider Prescription Habits for Folic Acid Prophylaxis

Graph 5: Provider Prescription Habits for Hydroxyurea Prophylaxis



While not part of the national guidelines, in an attempt to learn about additional provider habits for pediatric sickle cell disease patients we asked about the proportion of patients taking Ciklaviv. The majority of participants (15) estimated that <10% of their patients were taking

Ciklavit, 10 did not know, 2 estimated 11-25% and 1 estimated 51-75%. When asked about their prescribing habits of Ciklavit, 13 (41.9%) never did, 6 (19.3%) rarely did, 3 (9.7%) sometimes did, 3 (9.7%) found it unavailable, 1 (3.2%) always did and 5 (16.1%) did not answer. When asked why they did not always prescribe Ciklavit there were 20 responses, 8 (40%) of respondents were either unfamiliar with Ciklavit or unsure of its efficacy, 5 (25%) found the cost too high or the medication unavailable, 3 (15%) stated that it was not part of their protocol, 3 (15%) referred out to a specialist and 1 (5%) listed other. Lastly, when asked about herbal preparations for patients, 22 did not know if their patients were taking home remedies or herbal treatments, 2 estimated <10%, 1 estimated 11-25%, 1 estimated 26-50%, 1 estimated 51-75% and 4 did not answer. When asked to specify what their pediatric SCD patients were taking if they were taking herbal treatments, participants stated a leaf or herb mix was being used or that they did not know.

**Table 1: Provider Prescription Habits for SCD Complication Prophylaxis from Survey Sample of Nigerian Healthcare Providers, 2018 (Respondents N=31)**

	Always	Often	Sometimes	Rarely	Never	Not Available	No Response
Penicillin Prophylaxis	4	5	6	6	7	0	3
Malarial Prophylaxis	27	1	1	0	0	0	2
Pneumococcal Vaccination	9	5	6	3	3	2	3
Folic Acid Prophylaxis	29	0	0	0	0	0	2
Hydroxyurea Prophylaxis	0	6	8	6	4	4	3
Ciklavit	1	0	3	6	13	3	4

### 3.5 Section V: Furthering Education

When asked about interest in continuing education on sickle cell disease, 30 (96.8%) of participants said yes. When asked to check all the preferred method of education the top 3 were that 23 (74.2%) preferred E-learning modules, 21 (67.7%) online courses and 15 (48.4%) live educational sessions.

#### **4. Discussion**

Nigeria has the highest burden of sickle cell disease in the world, with an annual infant death rate of about 100,000 or 8% of all infant mortality in the country (Nigeria 2014). The burden of this disease has far reaching effects and there needs to be a more coordinated effort to standardize detection, diagnosis, prevention and treatment of sickle cell disease, especially in the under-five population. In an attempt to streamline care and offer a protocol for how to approach the prevention, diagnosis and treatment of sickle cell disease, the Nigerian government created the National Guideline for the Control and Management of Sickle Cell Disease in 2014. Since the creation of this guideline, there has not been a study to determine if the information that was meant to be used to provide a universal standard of care has been implemented into daily practice. In this study, we were able to gather information from various health providers across Nigeria on the awareness of the current practices outlined in the national guidelines, current treatment practices and determine possible barriers to care.

The national guideline is comprehensive and serves as a source for facing a variety of different issues related to sickle cell disease including management of acute complications, sickle cell disease and pregnancy, etc. We chose to focus our line of questioning on the routine prophylactic measures that are outlined in the national guidelines. For the purposes of prevention, we wanted to know what the clinic capabilities were to correctly diagnose sickle cell



disease patients to know when prophylactic treatment should be initiated. Prevention was also at the root of questions regarding clinical medical records and sickle cell disease databases to see the continuity of care in those who have sickle cell disease. The role of genetic counselors and newborn screening are also used in the national guidelines as key points for prevention of disease and/or initiation of prophylactic measures to prevent life threatening complications of sickle cell disease.

The core of the prophylactic measures center around malarial prophylaxis, prevention of anemia through folic acid supplementation, prevention of infections through penicillin prophylaxis and the addition of pneumococcal vaccinations to the immunization schedule. Additionally, hydroxyurea is cited as a prophylactic measure when indicated. While not listed in the national guidelines, we added the use of Ciklvait and knowledge of home remedies to gain a more holistic few of possible provider practices. Although our sample size is a small representation of the whole of the health providers in Nigeria, we were still able to determine some valuable gaps between the goals of the standard of care outlined in the national guideline and the actual practice that pediatric sickle cell patients are receiving.

The clinics that the participants work in were mostly public, urban teaching hospitals serving either pediatric specific or adult and pediatric patients. First, in a discussion of the different clinics that were represented in our participants, there was a range of different diagnostic methods that were available and that were most commonly used. In order to know if a patient requires specific treatment and prophylaxis for sickle cell disease, the disease status of the patient needs to be known. There national guidelines outline the availability and use of a sickling test, solubility test, hemoglobin electrophoresis, blood film, high performance liquid chromatography (HPLC) and isoelectric focusing (IEF) as possible diagnostic tools suitable for

sickle cell disease. When asked to mark all of the diagnostic options available at their facility, we found that 30 of the institutions had electrophoresis (cellulose acetate, citrate agar), 3 marked capillary electrophoresis, 9 marked HPLC and 1 marked IEF. The methods of diagnosis that were marked were 27 marking electrophoresis (cellulose acetate, citrate agar), 3 marked capillary electrophoresis, 3 marked HPLC and 1 marked that they did not know.

When it came to record keeping, all of the facilities were determined to medical records with roughly half of those keeping a database of sickle cell disease patients. Despite the encouragement for the use of genetic counselors in the national guidelines to aid in education about sickle cell disease and its route of inheritance, the majority of institutions did not have a genetic counselor available. Perhaps the most concerning, is that despite the strong push for newborn screening from a national level, 87.1% of the respondents indicated that newborn screening is not done at their facility. There might be some confounding variables since we did not gather information from OB-GYN specific centers, but pediatricians account for a large number of newborn screenings, especially for those who serve patients that may have been born at home. Newborn screening can aid in early detection and early prophylaxis or treatment to help decrease the rate of those who die from early complications of the disease.

In an effort to better understand the background of the study participants, there was a section specific to the provider demographics. The resulting information showed that most of the providers were pediatricians holding an MBBS degree, training as a fellow or an MD. When asked about their patient population, most of the respondents saw up to 10 pediatric sickle cell disease patients per week. Given the prevalence of the disease in the country and the lack of surveillance and newborn screening examinations, it was important to open up the survey to participants that were not just pediatric sickle cell disease specific providers. Since the disease is

so common, it is not just those who are specifically trained in pediatric sickle cell disease that need to be aware of the national guidelines and prophylactic measures, but all healthcare providers need the knowledge to know how to identify those patients with sickle cell disease, correctly diagnose and provide prophylactic care.

When it came to the knowledge of the national guidelines, there seemed to be a disconnect between the goals of the guidelines to influence provider practice and the dissemination of the guidelines so that they could be used. The national guidelines were published in November of 2014 and yet in March of 2018, almost four years later, half of the respondents had not heard of any set of national guidelines and 58.1% had not heard of the National Guideline for the Control and Management of Sickle Cell Disease (2014) in particular. This is a huge gap in awareness and something needs to be done to better disseminate this information throughout the Nigerian medical community, otherwise a standardized method of care cannot be accomplished. Given the lack of knowledge about the existence of the national guidelines, it is not surprising that the majority of the participants did not know if they were following them in their treatment of pediatric sickle cell disease patients. When questioned about why the national guidelines were not always followed, the reasons were most commonly cited as not knowing about the guidelines, not having a copy of the guidelines and not being the primary SCD provider. It was interesting to note, that of those providers who were pediatricians specifically, 12 were not aware of the national guidelines and 10 were aware. Given the focus of this study on pediatric sickle cell disease healthcare, the pediatricians responses were used to cross tabulate information between other types of providers and their knowledge of the national guidelines.

Despite the majority of providers not being aware of or using the national guidelines as their primary influence on pediatric sickle cell disease care, there were still some elements of practice

that fell into line with the goals of the standardized care outlined in the guidelines. For example, the areas of highest patient population treatment percentage and provider prescribing frequency were malarial prophylaxis and folic acid supplementation. Given the high rates of death due to complications of malaria and anemia in sickle cell disease patients, this is encouraging data.

However, given the complications of disease brought on by infection, it was surprising to see that penicillin prophylaxis and pneumococcal vaccination was still not always routinely seen in patients or prescribed. In the case of penicillin prophylaxis the most common themes that arose for not always prescribing penicillin prophylaxis for pediatric sickle cell disease treatment was that it was not a part of the institutions routine treatment protocol or that there was not enough evidence that the use of penicillin prophylaxis was helpful. For pneumococcal vaccinations, the most commonly cited reasons for not prescribing this for their pediatric sickle cell disease patients was due to concerns about cost or children already being immunized. Given that both penicillin prophylaxis and pneumococcal vaccinations are outlined as prophylactic measures in the national guidelines, more information and awareness about their role in preventing infection and complications needs to be conveyed.

Hydroxyurea treatment was another interesting difference between the standard of care outlined in the national guidelines and the responses from this survey. Given the different requirements to determine the need for hydroxyurea treatment depending on disease severity, there could have been some confusion when asked about prescription habits in a general pediatric sickle cell disease population. We found that 35.5% thought that <10% of patients were taking hydroxyurea, however 32.3% did not know. Given that hydroxyurea could have different implications on treatment and disease state, the amount of providers that did not know if their patients were taking this was concerning.

Lastly, we were curious to learn about the proportion of the patient population that was taking treatments that were not outlined in the national guidelines, but are used in actual practice. In the case of ciklavit, about half of the providers estimated <10% of their patients were taking ciklavit, but again there were 32.3% who did not know. When asked about their prescribing habits of ciklavit about half responded that they never prescribe it, while the others did sometimes or rarely. The cost and unfamiliarity with its treatment efficacy were cited as reasons for not always using it. Lastly, in an effort to better understand home remedies and herbal treatments, we wanted to know how many patients were taking these and what they were using if they were. Again, almost 71% of the respondents did not know and of those who did they weren't sure what the treatment was aside from it being a leaf or herb mix.

In concluding the study, there was a great interest in wanting to learn more about sickle cell disease through a variety of different online and in person educational methods. This lines up well with the recommended changes and implementations that could take place to better disseminate the information that is provided in the national guidelines. While there is still a long way to go for the standardized treatment of pediatric sickle cell disease in Nigeria to decrease the burden of disease and under-five mortality, health providers are willing and interested to improve treatment practices and patient care.

The limitations of this study largely fall on access to internet for survey completion since many medical professionals in Nigeria rely on the use of data on personal devices to access the internet. Another limitation could be bias due to convenience sampling. With the bulk of recruiting being done at the joint 14<sup>th</sup> Triennial Congress of Union of National African Paediatric Societies and Associations (UNAPSA) and the 49<sup>th</sup> Paediatric Association of Nigeria (PAN) Annual General Meeting and Scientific Conference in Abuja, Nigeria, that could target an

audience that is personally invested in being kept up to date on the most recent and well-established treatment options. Due to the technology availability and success of the online survey formation, there was a smaller respondent size than hoped for that could be difficult to form conclusions from. However, there was still a response rate of over 30% and while overt generalizations cannot be made, trends were still obtained and recommendations can be created. The results will help provide a foundation to guide future research projects.

## **5. Conclusion**

While the sample size of this study is small and not representative of all the healthcare providers in Nigeria, there are still some important conclusions that can be reached. The first goal of the study was to evaluate current provider awareness and knowledge of the National Guideline for the Control and Management of Sickle Cell Disease of 2014. The results of those who participated in this study showed that about half of the providers were not aware of the guidelines. Of the healthcare providers in this survey, 18 (58.1%) had not heard of the national guidelines, leading to the conclusion that there is a gap in the national standard and current practice if the majority of providers are not aware of what the national standard of care is. Given that this was a core healthcare push by the Nigerian government to standardize care and reduce the rate of under 5 child mortality, there is more that needs to be done. In an effort to increase the awareness of these guidelines and adherence to the recommended healthcare protocol there needs to be a better dissemination of information. Given the prevalence of the disease, these recommendations should be incorporated into medical training and daily practice.

The next major conclusion is there needs to be a more coordinated effort to implement prevention strategies for the complications of pediatric sickle cell disease. The two most

important steps for this would be a universal newborn screening program and the use of genetic counselors. Both of these are recommendations in the national guidelines, but that showed low utilization among the institutions of the study participants. The use of these would provide the basis for earlier detection and prophylactic measures to reduce the rates of preventable mortality. In terms of prophylactic treatment, malarial prophylaxis and folic acid supplementation showed good utilization among all the participants. However, infection prevention with the penicillin prophylaxis, pneumococcal vaccination and hydroxyurea were still inconsistent and not standardized. Other treatment practices like ciklavit and herbal medications were largely either unused or used without direct provider knowledge.

Lastly, there was high participant interest in furthering their sickle cell disease knowledge and a desire to learn more. This provides hope for the future and for those who have sickle cell disease in Nigeria. While the current rates of pediatric sickle cell disease and associated under-5 mortality are still high, there is growing awareness and interest in medical providers to combat this disease burden. With the creation of the National Guideline for the Control and Management of Sickle Cell Disease, the goal for care is set and the providers want to improve care. In order for these guidelines to be implemented and the providers to follow these standards, there needs to be work done to bridge the gap and to make this information more accessible and utilized throughout the country. Important steps have been taken, but only when the protocol moves to implementation can we hope to see the necessary changes for Nigeria optimally manage its pediatric sickle cell disease burden.

## 6. Reflection

In concluding this project there were a number of important points to reflect on. First, was the realization of all the work that goes into conducting research. I have never been involved in the research aspect of public health and medicine before, so this was a very eye-opening experience. There is so much careful planning and prep work that occurs even before the initiation of the project that was interesting to learn about and to be a part of. The second realization that I had was the vast opportunities available in the realm of global health. I had committee members that were physicians, researchers and public health specialists and each person plays an integral role in the field of global health as a whole. As a student with the hopes of going into global health, this was encouraging and exciting. Lastly, I realized how much I am capable of as an MD/MPH student. This was the first time that I have been able to apply my knowledge and my passions in a substantial way that can help contribute to the field of work that I have always wanted to be a part of.

The global health community at UNMC is far reaching and is doing amazing and innovative work to help make it a better place for all. I think that being able to complete my MPH and to have worked collaboratively with people that are doing these amazing things to complete a project that may serve to help others is very fulfilling. I know that the experiences that I have had throughout this process have helped to shape my perspective and my understanding of the public health world, which will also ultimately play a large role in who I become as a physician. There is still much work to be done in both the field of medicine and public health in both local and international contexts, but I feel equipped and excited about the part that I might play to help those who are in need.



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**Appendix A:**

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**Health Professional Treatment Practices for Sickle Cell Disease in Nigeria**

Please complete the survey below.

Thank you!

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02/27/2018 1:02pm [www.projectredcap.org](http://www.projectredcap.org) 

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Nigeria has the largest burden of sickle cell disease worldwide and this disease greatly impacts the health care of this country. This study will explore the differences in sickle cell disease treatment and knowledge between different types of practitioners and different treatment settings. Information gathered from this study will describe the current knowledge base, factors impacting health care decisions, what is currently being done for treatment and where improvements can be made to better address this health burden.

- Yes  
 No

You are being asked to participate in this study because you are a practicing health care professional in Nigeria. The data taken from this study may be used to help inform treatment guidelines and practices for sickle cell patients.

This study is being administered by the International Foundation Against Infectious Disease in Nigeria (IFAIN) and the University of Nebraska Medical Center (UNMC). Participation in this study is completely voluntary and will consist of answering a selection of online survey questions regarding your treatment practices for sickle cell disease patients. The questions will range from where your treatment information comes from to the treatment options most likely used. I do not anticipate any risks to you participating in this study other than those encountered in daily life. All information gathered will be treated with strict confidentiality. We will protect information about your participation in this research to the best of our ability. You will not be named in any reports or publications and all of your information will be coded with a unique identification number following completion. Only approved study personnel will have access to your information. Following completion of the survey you will receive 3000Naira in data credit sent to the mobile number on file to compensate you for your time and data usage.

If you have any questions, this study is being conducted by MDIMPH student Natalie Wichelt and Dr. Stephen Obaro. Any questions or concerns can be sent to Natalie Wichelt at [natalie.wichelt@unmc.edu](mailto:natalie.wichelt@unmc.edu) or Dr. Obaro at [Stephen.obaro@unmc.edu](mailto:Stephen.obaro@unmc.edu). If you have any concerns regarding your rights as a subject in this study you may contact the Institutional Review Board at UNMC (402)-559-6463; [IRBORA@unmc.edu](mailto:IRBORA@unmc.edu).

Statement of Consent: I have read the above information and by continuing with the survey consent to participating in this study.

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**Section I: Clinic Characteristics**

What best describes your place of primary practice?

- Emergency Room/Department  
 Hospital Inpatient  
 Hospital Outpatient  
 Teaching Hospital  
 General Clinic  
 Specialized Sickle Cell Disease Center  
 Primary Healthcare Center  
 Other

If other, please specify \_\_\_\_\_

What best characterizes the institution that you work in? (check all that apply)

- Adult  
 Pediatric  
 Both Adult and Pediatric  
 Private  
 Public

Is your institution considered urban or rural?

- Urban  
 Rural

Does your primary place of practice keep medical records?

- Yes  
 No  
 I don't know

Does your treatment center keep a database of Sickle Cell Disease patients?

- Yes  
 No  
 I don't know

What equipment is available at your facility? (check all that apply)

- CT  
 MRI  
 Transcranial Doppler Ultrasounds  
 X-Ray/Radiography  
 I don't know  
 None  
 Other

If other, please specify \_\_\_\_\_

What are the methods available for diagnosis of Sickle Cell Disease at your facility? (check all that apply)

- Capillary Electrophoresis  
 Electrophoresis (cellulose acetate, citrate agar)  
 High-Performance Liquid Chromatography (HPLC)  
 Isoelectric Focusing (IEF)  
 I don't know  
 Other

If other, please specify \_\_\_\_\_

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Which method of diagnosis is most commonly used to diagnose Sickle Cell Disease at your facility?

- Capillary Electrophoresis
- Electrophoresis (cellulose acetate, citrate agar)
- High-Performance Liquid Chromatography (HPLC)
- Isoelectric Focusing (IEF)
- I don't know
- Other

If other, please specify

\_\_\_\_\_

Is there a genetic counselor available at your institution?

- Yes
- No
- I don't know

Is newborn screening done at your facility?

- Yes
- No
- I don't know

**Section II: Provider Demographics**

What is your gender?

- Male
- Female
- Choose not to disclose

What is your age? (in years)

\_\_\_\_\_

How many years have you been in practice? (in years)

\_\_\_\_\_

Please mark the highest level of education you have completed. (check all that apply)

- MBBS
- MD
- PhD
- Masters
- MPH
- RN
- Other

If other, please specify

\_\_\_\_\_

From which institution did you receive your most recent degree?

\_\_\_\_\_

Please mark the option that best describes your position?

- Pediatrician
- General Practice
- Pediatric Resident/Registrar
- Pediatric Subspecialist not Hematologist
- Medical Officer
- Nurse
- Community Health Worker
- Other

If other, please specify

\_\_\_\_\_

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In which state of Nigeria do you practice?

\_\_\_\_\_

On average, how many patients do you see per week?

\_\_\_\_\_

On average, how many of the patients you see in one week are pediatric?

\_\_\_\_\_

On average, how many of the pediatric patients you see in one week have sickle cell disease?

\_\_\_\_\_

**Section III: Guideline Knowledge**

4. Are you aware of a Nigerian National Guideline for Sickle Cell Disease treatment?  Yes  No

Have you heard of the NATIONAL GUIDELINE FOR THE CONTROL AND MANAGEMENT OF SICKLE CELL DISEASE (2014)?  Yes  No

Do you follow the national guidelines for Sickle Cell Disease treatment?

	Always	Often	Sometimes	Rarely	Never	I don't know
	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

If you do not always follow the national guidelines for Sickle Cell Disease treatment, why? (please check all that apply)

- I did not know there were national guidelines
- I do not have a copy of the national guidelines
- I prefer to use other guidelines
- Medications listed in the guidelines are not available where I work
- Vaccines listed in the guidelines are not available where I work
- Labs recommended in the guidelines are not available where I work
- I am not the primary Sickle Cell Disease patient provider
- I have not received Sickle Cell Disease patient care specific training
- Other

If you marked Other, please list up to 5 reasons why you do not follow the national guidelines.

\_\_\_\_\_

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**Section IV: Treatment Practices**

Of the pediatric patients with Sickle Cell Disease you have seen, what proportion are taking penicillin prophylaxis?  
(in a percentage, %)  
(can write "I don't know" in the box)

	Always	Often	Sometimes	Rarely	Never	Penicillin is not available	I do not have the ability to prescribe
How often do you prescribe penicillin prophylaxis for your pediatric Sickle Cell Disease patients?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

If you do not always prescribe penicillin prophylaxis, why?

Of the pediatric patients with Sickle Cell Disease you have seen, what proportion are taking malarial prophylaxis?  
(in a percentage, %)  
(can write "I don't know" in the box)

	Always	Often	Sometimes	Rarely	Never	Malarial Prophylaxis Not Available	I do not have the ability to prescribe
How often do you prescribe malarial prophylaxis for your pediatric Sickle Cell Disease patients?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

If you do not always prescribe malarial prophylaxis, why?

Of the pediatric patients with Sickle Cell Disease you have seen, what proportion have received a pneumococcal vaccination?  
(in a percentage, %)  
(can write "I don't know" in the box)



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	Always	Often	Sometimes	Rarely	Never	Pneumococcal Vaccine Not Available	I do not have the ability to prescribe
How often do you prescribe pneumococcal vaccinations for your pediatric Sickle Cell Disease patients?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

If you do not always prescribe pneumococcal vaccines, why?

---

Of the pediatric patients with Sickle Cell Disease you have seen, what proportion are taking folic acid? (in a percentage, %)  
(can write "I don't know" in the box)

---

	Always	Often	Sometimes	Rarely	Never	Folic Acid Not Available	I do not have the ability to prescribe
How often do you prescribe folic acid for you pediatric Sickle Cell Disease patients?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

If you do not always prescribe folic acid, why?

---

Of the pediatric patients with Sickle Cell Disease you have seen, what proportion are taking hydroxyurea? (in a percentage, %)  
(can write "I don't know" in the box)

---

	Always	Often	Sometimes	Rarely	Never	Hydroxyurea Not Available	I do not have the ability to prescribe
How often do you prescribe hydroxyurea for your pediatric Sickle Cell Disease patients?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

If you do not always prescribe hydroxyurea, why?

---

Of the pediatric patients with Sickle Cell Disease you have seen, what proportion are taking ciklavit? (in a percentage, %)  
(can write "I don't know" in the box)

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	Always	Often	Sometimes	Rarely	Never	Ciklavit Not Available	I do not have the ability to prescribe
How often do you prescribe ciklavit for you pediatric Sickle Cell Disease patients?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

If you do not always prescribe ciklavit, why?  
\_\_\_\_\_

Of the pediatric patients with Sickle Cell Disease you have seen, what proportion are taking home remedies or herbal treatments?  
(in a percentage, %)  
(can write "I don't know" in the box)  
\_\_\_\_\_

If your pediatric Sickle Cell Disease patients are taking herbal treatments, please specify what?  
\_\_\_\_\_

**Section V: Furthering Education**

Would you be interested in continuing education on Sickle Cell Disease?  
 Yes  
 No  
 I don't know

Which type of educational format would you prefer? (check all that apply)  
 Educational Pamphlets  
 E-learning Modules  
 Educational Surveys  
 Live Educational Sessions  
 Webinar  
 Online Courses

Can we keep your e-mail for surveys in the future?  
 Yes  
 No

Please provide your mobile phone number for compensation purposes only. 3000Naira in data credit will be sent to the mobile phone number provided once completion of the survey has been confirmed.  
\_\_\_\_\_

