African Chronicles Research Publication Journals. p-ISSN 3121-8989, e-ISSN 3121-8970
Okwe et al (2024). Afr Chron Res Pub J; 1(1): 70-87
Article URL: https://researchpubjournals.org/?p=1320
Public Health Research

Research proposal on the perception of sickle cell patients on their diagnosis and management practices at the eighteen sickle cell clinics of Delta State Nigeria

Okwe UN[1,2], Nwose EU[1,3], Ofili CC[1]

[1]Department of Public and Community Health, Novena University, Ogume, Nigeria
[2]Delta State Ministry of Health, Asaba. Nigeria
[3]School of Health and Medical Sciences, UniSQ, Toowoomba Australia

Corresponding Author:
Dr. Uche Okwe. Delta State Ministry of Health, Asaba. Nigeria. Email: tansinwanney@yahoo.com, Tel: +2348037615027

Received date: 27 August 2024
Revised date: 29 October 2024
Accept date: 03 November 2024

African Chronicles Research Publication Journals. p-ISSN 3121-8989, e-ISSN 3121-8970
Okwe et al (2024). Afr Chron Res Pub J; 1(1): 70-87
Article URL: https://researchpubjournals.org/?p=1320
Public Health Research

Abstract

Background: Sickle cell disease is an autosomal recessive. Prevalence depends on the sickle cell trait. Nigeria is the most Sickle Cell Disease endemic country globally. This is a serious Public Health issue in the 21st century and a challenge to health care system in Delta State where there are 11,000 SCD patients registered in 24 SCD clinics in the state. Nigeria has a significant prevalence of SCD trait making strategic resource allocation for disease a crucial priority.

Objective: To investigate the perceptions of sickle cell patients on their diagnosis and management practices at the eighteen SCD clinics in the 3 senatorial districts of the state.

Methods: Study is cross- sectional, descriptive, mixed-methods study. Measures of knowledge and attitude of SCD patients, effectiveness of management, and challenges of healthcare system would be assessed among 700 participants randomly -selected from 18 sickle cell clinics across 3 senatorial districts of Delta State. Qualitative and qualitative data would be collected using semi-structured interviews involving 60 randomly selected individual teenagers among SCD patients. There will be a 10-months' retrospective clinical trial using randomly -selected clinics on hydroxyurea usage in crisis management. Timeline for the whole study is 30 months.

Expected outcome: The results of the study will show empirical data on various components of Behavioural changes wheel on the management and healthcare system in Delta SCD clinics.

Significance: The outcome of this study will hopefully improve the quality and sensitization of health professionals, policy makers, and resource allocators in Delta State which is a very vital step in making informed decisions on the management and control of SCD.

Keywords: SCD, Perception, Diagnosis, Management Practices, Sickle cell Clinics

African Chronicles Research Publication Journals. p-ISSN 3121-8989, e-ISSN 3121-8970
Okwe et al (2024). Afr Chron Res Pub J; 1(1): 70-87
Article URL: https://researchpubjournals.org/?p=1320
Public Health Research

1.Background

1.1. Introduction

Nigeria has a population of 232.6 million with an annual growth rate of 2.12% (United Nations, 2024). Sickle Cell Disease (SCD) is a significant public health concern in Nigeria with a remarkable high carrier prevalence of approximately 25% for the AS genotype (Modell, 2008) while the AC trait predominantly found among the Yoruba people of southwestern Nigeria affecting about 6% of this population (Balogun et al., 2010). With a higher black population, it bears the greatest burden of SCD (Adewoyin, 2015). Out of an estimated 5.4 million expected live births in 1988, approximately 90,000 newborns are expected to have SCD while about 1.1 million will carry the sickle cell trait (AS). The clinical phenotype of sickle cell anemia (SS) is severe with manifestations occurring very early in childhood with mean Hemoglobin level 7.6 g/dl and fetal hemoglobin 5.9g/dl. A very high infant mortality due to infections occurs especially in rural areas Akinyanju (1989).

Knowledge regarding SCD is needed by the carriers of SCD trait to enable informed decisions about their reproductive life and health related choices so that the burden of social stigma by sickle cell sufferers could reduce (Adewoyin, 2015). Nigeria's lack of adequate support structures worsens the plight of clinically depressed sickle cell patients who already face stigma, discrimination, and social exclusion (Ezenwosu et al., 2021; Salih, 2019). In Delta State, despite the sickle cell prevalence and control activities, awareness remains low in the rural areas, with mostly health professionals and community initiatives indicating a need for enhanced public education and awareness campaign within the health care system (Awunor & Erusiafe, 2016; Uchechukwu, 2016).

The limited awareness of sickle cell causes, symptoms and prevention among patients, underscores a significant knowledge gap (Adigwe, 2022), potentially compromising effective disease management and control strategies (Tusuubira et al., 2019). With the resolution declaring SCD a global public concern by the United Nations General Assembly in 2009 (Mulumba &Wilson, 2015), Delta State Government and Non -Governmental Organizations have prioritized the development of comprehensive SCD health system frame work by allocating annual funds for universal coverage of SCD patient, budgeting for control activities, training genetic counsellors , upgrading the sickle clinics in 23 LGAs and a referral centre (Asaba Metro news, 2022) and fostering community and family participation in SCD

management to enhance knowledge, awareness and effective management (see SCD health system framework is illustrated in figure 1).

1.2. Literature Review

Sickle cell disease has been recognized for more than 100 years (Mahran et. al., 2019). Globally, 330, 000 babies are born with major hemoglobinopathy out of which 275,000 have SCD (Aygun & Odame, 2012; Piel et al., 2013). United Nations estimates that 20 to 25 million people worldwide live with SCD of which 12-15 million live in Africa (Adigwe et al., 2023). It has been estimated that 75% to 85% of children born with SCD are in Africa, where mortality rate for the under-5 is 50% to 80% (Makani et al., 2011; Aygun & Odame, 2012).This is because of poverty, inadequate number of health facilities or clinics, inaccessibility of hospitals in rural areas, cost of treatment, poor hematology services, reduction in number of health workers and care givers, inadequate training on SCD and its management and late detection of SCD (Odame, 2010). Morbidity affects all organs of the body. There is painful crisis, avascular necrosis of the musculoskeletal system (Balogun et al., 2010), severe anemia (Bello-Manger et al., 2020), stroke (Kolapo et al., 2011), sepsis and chronic illness which affect quality of life and can lead to death. In Nigeria, many births and deaths occur with traditional birth attendants and faith- based organizations and are not recorded (Olatunya et al.,2021).

This limits the statistical data on the incidence and deaths of SCD patients in Africa (Serjeant, 2010). The Nigerian Government on its part has taken some measures towards providing quality healthcare services for SCD such as the development of the National Guideline for the Management and Control of SCD (Ibemere et al., 2023), establishment of six zonal Centres of Excellence for SCD equipped with HPLC for early detection and comprehensive care, creation of a National Desk for SCD at the Federal Ministry of Health (FMoH) and more. Clinical care is available for patients with SCD mainly in the secondary and tertiary healthcare facilities with hydroxyurea use as first line of treatment in SCD crisis (Okocha et al., 2013), but most patients with SCD remain unreached in the rural communities of Nigeria (Isa et al.,2023). In Delta State, there has been upgrading and equipping of 23 sickle cell clinics and construction of a referral centre after which the State House of Assembly in May 2023 passed into law the establishment of 24 centres for collection, coordination and distribution of data on sickle cell and mandatory newborn hemoglobinopathy screening (Asaba Metro news, 2023; The Guardian news, 2023).

African Chronicles Research Publication Journals. p-ISSN 3121-8989, e-ISSN 3121-8970
Okwe et al (2024). Afr Chron Res Pub J; 1(1): 70-87
Article URL: https://researchpubjournals.org/?p=1320
Public Health Research

Universal health coverage has also been introduced (Ajala-Damisa & Agbaoye 2021; Ochei, 2024). This was done to increase awareness of the people living with sickle cell and ensure appropriate management of the disease as soon as babies are diagnosed with SCD and when SCD patients access treatment. To enable control strategies yield significant results, there is a need to improve knowledge about SCD by health education, health care financing, genotype testing, counselling, registration at the SCD clinics to enable effective management among urban and rural communities in Delta State (Akinyanju, 2010; Awunor & Erusiafe, 2016). This goal is conceptualized in Delta State SCD health system framework (DHPRS SMOH, 2015) shown in figure 1.

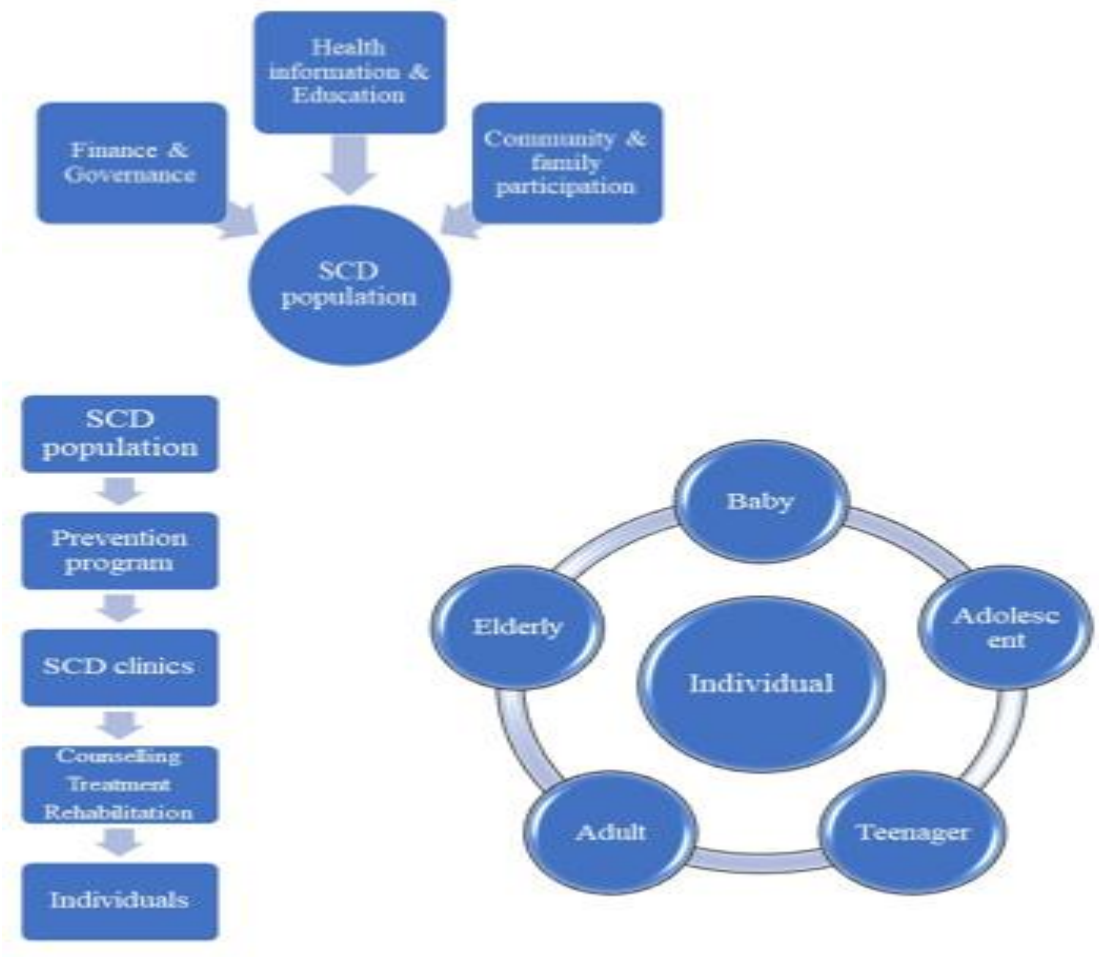

Fig 1: Delta State Health system framework of Delta State Ministry of Health, 2015

1.3. Statements of the Problem

i. Most SCD crisis recorded are readily amenable to treatment especially in Delta state, but the interventions and treatments are scarce and if available not accessed by majority

African Chronicles Research Publication Journals. p-ISSN 3121-8989, e-ISSN 3121-8970
Okwe et al (2024). Afr Chron Res Pub J; 1(1): 70-87
Article URL: https://researchpubjournals.org/?p=1320
Public Health Research

of the patients that live in low resource settings (Efe, 2013; Odunvbun et al., 2008, Okocha et al., 2022).

ii. In Delta State , there is inadequate human and financial resources of health (Efe, 2013) to effect care at the established sickle cell clinics.

iii. In Nigeria the number of sickle cell clinics are inadequate (Adigwe et al., 2023; Collier, 2012; Galadanci et al., 2014).

iv. Persons with SCD often face stigma (Leger et al., 2018) which interferes with their psychosocial development, education and work opportunities (Moussa et al., 2023).

v. There is no documented evidence to show the perception of the Delta State SCD patients towards their clinical condition, psychosocial development and management practices introduced and Implemented at the sickle cell clinics of Delta State. This justifies why this research needs to be conducted in Delta State.

1.4. Broad Objective

The aim of this research is to investigate the "Perception of the Sickle cell patients towards their diagnosis and efficacy of management practices at the Sickle cell disease clinics of Delta State."

1.4.1. Specific objectives

i. To evaluate the knowledge and attitude of the SCD patient towards information regarding his diagnosis.

ii. To evaluate the level of care and efficacy of management of sickle cell disease.

iii. To identify gaps and challenges in our health system that may cause an increase in the mortality of SCD.

iv. To assess the effectiveness of established use for prevention of SCD crisis in Delta State Sickle cell clinics.

2. Methods

2.1. Research design

The proposed study design is mixed methods. Table 1 provides a summary of the different design for each specific objective. The proposed methods would include a cross-sectional, descriptive approach integrating both qualitative and quantitative approaches. Measures of knowledge and attitude of SCD patients, effectiveness of management, and challenges of healthcare system would be assessed among 700 participants.

There will be a cohort study i.e. longitudinal design component. This would involve 10-months' retrospective clinical audit to analyse medical records from randomly selected SCD clinics to assess hydroxyurea effectiveness in managing sickle cell crisis. Hence timeline for the whole study is estimated to be about 30 months.

African Chronicles Research Publication Journals. p-ISSN 3121-8989, e-ISSN 3121-8970
Okwe et al (2024). Afr Chron Res Pub J; 1(1): 70-87
Article URL: https://researchpubjournals.org/?p=1320
Public Health Research

Table 1: shows study specific objectives, research questions, hypothesis, and design

Specific objectives	Research questions	Hypothesis	Study design
To evaluate the perspective of the SCD patient towards his/her management practices	Do the SCD Patient in Delta have a perspective of SCD management Practice?	There is no significant association between the SCD client age and his/her perceptive and management practice.	Cross-sectional, descriptive, quantitative analysis
To evaluate the level of care and efficacy of management of SCD	Do the SCD Patient benefit from efficient health care service delivery?	There is no significant association between the benefit of the SCD and efficient health care service delivery at the clinics	Cross-sectional, descriptive, qualitative, quantitative analysis
To identify the gaps and challenges in Delta State Health System	Are there gaps/challenges in the health system that affects SCD?	There is no significant association between challenges and gaps in our health system that affect SCD management	Exploratory (Qualitative analysis)
To assess the effectiveness of the established use of hydroxyurea for prevention of SCD crisis in our clinics	Is the already established mechanism of SCD crisis management with hydroxyurea effective?	There is no significant association between the established mechanism of prevention of SCD crisis with hydroxyurea and management of SCD crisis	Quantitative analysis

2.2. Data collection and sample size distribution

As illustrated in table 1, the research methodology is guided by an outline and overview of the research framework objectives, questions, hypothesis, and study design. Table 2 represents the distribution of clients selected from the sickle cell clinics in the State districts participating in this research study.

Sampling technique would include random selection among a total of 11,000 SCD patients registered in 24 SCD clinics in the state (DHPRS Records, 2023). A total of 18 clinics will be randomly selected among 24 SCD clinics in 3 senatorial districts of Delta State (DHPRS Records, 2023). The selection criteria is as defined on table 3.

Table 2: Sickle cell clinics and clients selected by state districts

State districts	SCD clinics	No of clients
Delta Central	Sapele, Ughelli, Otujeremi, Eku, Oghara, Orerokpe	233
Delta North	Agbor, Ogwashi-uku, Asaba, Kwale, Secretariat, Okwe	234

Delta South	Warri, NPA, Oleh, koko, Burutu, Bomadi	233
	TOTAL	**700**

Table 3: shows the inclusion and exclusion criteria of selecting SCD participants

S/No	Inclusion	Exclusion
1.	All sickle cell patients	Parents not allowed to participate in this study
2.	Those that voluntarily choose to participate	Those not willing to participate
3.	Those that are well during data collection	SCD patients that are ill during data collection
4.	Sickle cell clinics randomly selected are to participate	Health care workers in the sickle cell clinics are not allowed to fill questionnaires

2.3. Scope of the Research

The scope of this research encompasses an investigation of the perception on the management of SCD amongst sickle cell clients in the Delta State sickle cell clinics that are located within in the three state senatorial districts.

2.4. Operational process/Achievability

Three research assistants from the Department of Program, Research and statistics of the Ministry of Health who have been trained on program monitoring and evaluation will be recruited and trained on the modalities of questionnaire administration and collection. The semi -structured confidential interviews are to be done by the author who will visit the SCD clinics on their clinic days. Questionnaires will be checked for proper completion on collection from participants. Data will be coded and kept confidential. A P-value of less than 0.05 will be considered statistically significant. Daily checking of filled questionnaires will be done at the end of each field day, to avoid incomplete data collection and ensure accuracy of data. For retrospective exploratory trial on use of hydroxyurea and health outcomes of the SCD patient, a retrospective cohort study with data sources from medical charts, SCD registry, hospital databases, hydroxyurea prescriptions and dosages, blood counts, renal function results, hospitalization and outpatient visit records, SCD diagnosis and severity (evidence) will be reviewed. Retrospective chart review form and questionnaires will be used for outcome assessment.

2.5. Statistics rationale/plan

2.5.1 Statistics/Justification**:**

1. Sampling: Stratified random sampling of clinics from each senatorial district to reduce sampling bias
2. Participant selection: Random sampling of 700 sickle cell participants from selected clinics to represent the population registered (11,000) at the SCD clinics. This will enable participants have an equal chance of being chosen.
3. Data collection: Administering questionnaires to 700 SCD patients. Semi structured–interviews will be done with 60 randomly- selected SCD teenagers of 13-19 years, 20 in each senatorial district (10 females and 10 males). This will be done in a sequential way with 15minutes' in-depth interviews culminating in a pilot -questionnaire study. Teenagers are chosen because of their outspokenness in expressing emotional and behavioral clues to questions on challenges of health care delivery at the sickle cell clinics and their active participation at the SCD clinics.
4. Will conduct a retrospective cohort exploratory trial with review of medical records of SCD patients for 10 months on hydroxyurea use patterns and outcomes in crisis management of sickle cell. This design is to help evaluate the effectiveness of hydroxyurea in real world settings using existing data.
5. Hypothesis testing: formulate null and alternative hypothesis with chi-square tests
6. Data analysis methods using tables, pie charts and histograms will be done. Statistics will be represented in simple percentages, cross tabulation, comparing column proportions in cross tabulations and chi-squared tests to enable data management and clarity of the results
7. Significance level will be set at 0.05 for hypothesis testing
8. Delta State SCD health system frame work is to explain why the research question is relevant.
9. Literature review is to summarize literature supporting the research hypothesis
10. Statistical analysis will be descriptive and inferential at 95% confidence level using the statistical package for social sciences (SPSS Inc. Chicago 11.) computer software version 22.0 to manage data efficiently
11. Research is significant as it will highlight challenges the SCD patients face in Delta State sickle cell clinics despite available resources.

African Chronicles Research Publication Journals. p-ISSN 3121-8989, e-ISSN 3121-8970
Okwe et al (2024). Afr Chron Res Pub J; 1(1): 70-87
Article URL: https://researchpubjournals.org/?p=1320
Public Health Research

2.6. SWOT analysis

To contextualize the research findings a SWOT analysis was conducted to identify the strengths, weaknesses, opportunities and threats related to sickle cell disease management in the state districts (Table 4).

Table 4: SWOT analysis

Strengths	Weakness	Opportunities	Threats
Established SCD clinics	Limited accessibility to far- to -reach areas of Delta State (rural and riverine	Increasing focus on interventions	Limited funding due to competing health programs
Growing interest in research of SCD	Stigma and misconceptions on SCD management	Collaborations with NGOs	High costs associated with SCD management
Support by Delta State ministry of health and NGOs	Maintaining participant engagement throughout the study	Increased funding for training of genetic counsellors	Difficulty in accessing patients' medical records
Literature reviews and research on SCD	Cost of travelling to various SCD clinics in all the LGAs is high	SCD care framework	Ethical concerns and permission to access records of patients

The validity of the questionnaire will be established through face and content validity criteria. Content validity measure will be ensured with the use of relevant literatures (Rodenberg, 2009) and (Feinstein, 1987). The questionnaire, based on objectives of the studies and Literature search will be given to experts in the field of medicine, research supervisory team made up of research supervisor, statistician and senior researchers in related field for thorough scrutiny. It will be checked for content, clarity, scope, appropriateness, and ability to elicit accurate information with respect to research objectives, questions and hypotheses (Masuwai et al., 2024). The reliability will be ensured using a test-retest approach (Zaki et al., 2012) using 10% of the sample size administered in a Sickle cell clinic different from the one chosen from the study. The internal consistency reliability will be calculated using Cronbach's alpha (Vaske et al., 2015). The normal range of values for coefficient alpha is between 0.01 and + 1.00. The reliability coefficient of 0.06 or above will be approved acceptable.

3. Expected Outcomes

African Chronicles Research Publication Journals. p-ISSN 3121-8989, e-ISSN 3121-8970
Okwe et al (2024). Afr Chron Res Pub J; 1(1): 70-87
Article URL: https://researchpubjournals.org/?p=1320
Public Health Research

3.1. Expected outcome

While extensive research has focused on perspectives of health care providers, there is knowledge gap regarding the health- related quality of life of SCD patients and effective management programs to enhance their well-being (Amaeshi et al., 2022). Most studies have prioritized health care professionals' views, leaving a critical need for patient-centered approach to better understand their experiences, identify gaps in current management programs and develop effective interventions. Despite Government initiatives aimed at mitigating SCD challenges in Delta State (Asaba Metro news, 2023; The Guardian news, 2023), many patients are dissatisfied with the quality of care received from primary health care providers (Aljuburi et al., 2012). The expected outcomes of this study will ensure policy makers understand the perspective of SCD patients on their diagnosis experience, assess patients' satisfaction on the physical, social, mental, psychological and spiritual health (Isa et al., 2023 & Nwagha et al., 2020), current management practices at Delta State SCD clinics and identify the challenges faced by patients in accessing quality care. The views and recommendations of the SCD patients on patient education, counselling, clinical and support group services, capacity building among health workers to enable behavioral change in issues concerning SCD (Campbell et al., 2023) will be incorporated to inform the study's findings and conclusion.

3.2. Significance of study

The study will contribute significantly to Delta State SCD management, equipping health care professionals, policy makers and stakeholders with contextualized solutions to improve incidence and treatment outcomes and inform evidence- based decision making.

The study will strengthen advocacy messages and advocacy tool kits that can be better designed to target social and cultural norms that have impeded early diagnosis and registration into the SCD clinics for better management.

4. Conclusion

The study highlights the relevance of improving the knowledge and awareness of Nigerians and particularly Delta State citizens in improving their perceptions to SCD management in the sickle cell clinics. It helps in understanding the causes and effective management of sickle ell disease. It also brings to light that the effective management of SCD is a multi-faceted approach incorporating medical, funding, health education, genetic counselling, genetic screening,

information sharing, community, and family approach. The findings underscore the need for access to health care services for a better health outcome for individuals with sickle cell disease.

4.1. Time frame:

This research from initiation till its complete will take 30 months.

4.2. Ethical Consideration

Ethical consideration and clearance will be gotten from the Novena University, Ogume to initiate the study and Delta State Ministry of Health, Ethical Committee (MOHREC) Asaba. Application for permission will be gotten from the individual hospitals to enable access to the sickle cell clinics domiciled within the hospitals**.**

Acknowledgements

Novena University, Ogume. Delta State; and Delta State Ministry of Health, Asaba. Nigeria

Conflict of Interest: nil

Data repository: Department of Public and Community Health, Novena University, Ogume. Delta State Ministry of Health

5. References

Abioye-Kuteyi, E. A., Oyegbade, O., Bello, I., & Osakwe, C. (2009). Sickle cell knowledge, premarital screening and marital decisions among local government workers in Ile-Ife, Nigeria. *African Journal of Primary Health Care and Family Medicine*,*1*(1). https://doi.org/10.4102/phcfm.v1i1.22

Adigwe, O. P., Onavbavba, G., & Onoja, S. O. (2023). Impact of Sickle Cell Disease on Affected Individuals in Nigeria: A Critical Review. *International Journal of General Medicine*, *Volume 16*. https://doi.org/10.2147/ijgm.s410015

Adewoyin, A. S. (2015). Management of sickle cell disease: A review for physician education in Nigeria (Sub-Saharan Africa). In *Anemia* (Vol. 2015). https://doi.org/10.1155/2015/791498

Agasa, B., Bosunga, K., Opara, A., Tshilumba, K., Dupont, E., Vertongen, F., Cotton, F., & Gulbis, B. (2010). Prevalence of sickle cell disease in a northeastern region of the Democratic Republic of Congo: What impact on transfusion policy? *Transfusion Medicine*, *20*(1). https://doi.org/10.1111/j.1365-3148.2009.00943.x

Ajala-Damisa, D., & Agbaoye, K. (2021). Accelerating progress towards UHC in Nigeria: The Delta State example. *Nigeria Health Watch [Online]*. https://articles.nigeriahealthwatch.com/accelerating-progress-towards-uhc-in-nigeria-the-delta-state-example/

Akinyanju, O.O. (1989) A Profile of Sickle Cell Disease in Nigeria. *Ann N Y Acad Sci* 565(1), 126-136. http://dx.doi.org/10.1111/j.1749-6632.1989.tb24159.x

Aljuburi, G., Phekoo, K. J., Okoye, N. O., Anie, K., Green, S. A., Nkohkwo, A., Ojeer, P., Ndive, C., Banarsee, R., Oni, L., & Majeed, A. (2012). Patients' views on improving sickle cell disease management in primary care: focus group discussion. *JRSM Short Reports*, *3*(12). https://doi.org/10.1258/shorts.2012.011173

Amaeshi, L., Kalejaiye, O. O., Ogamba, C. F., Adelekan Popoola, F., Adelabu, Y. A., Ikwuegbuenyi, C. A., Nwankwo, I. B., Adeniran, O., Imeh, M., & Kehinde, M. O. (2022). Health-Related Quality of Life Among Patients With Sickle Cell Disease in an Adult Hematology Clinic in a Tertiary Hospital in Lagos, Nigeria. *Cureus*. https://doi.org/10.7759/cureus.21377

Asaba Metro news (2022). World Sickle Cell Day: Dame Okowa inaugurates 23 sickle cell clinics. *Asaba Metro.* July 21st Edition. https://www.asabametro.com/tag/sickle-cell/

Aygun, B., Odame, I. (2012). A global perspective on sickle cell disease. *Pediatr Blood and Cancer*.59(2):386-390. https://doi.org/10.1002/pbc.24175

Balogun RA., Obalum, D.C., Giwa, S.O., Adekoya-Cole, T.O., Ogo, C.N., & Enweluzo, G.O. (2010). Spectrum of musculo-skeletal disorders in sickle cell disease in Lagos, Nigeria. *J Orthop Surg Res*, *5*, 2. https://doi.org/10.1186/1749-799X-5-2

Bello-Manga, H., Galadanci, A. A., Abdullahi, S., Ali, S., Jibir, B., Gambo, S., Haliru, L., Jordan, L. C., Aliyu, M. H., Rodeghier, M., Kassim, A. A., DeBaun, M. R., & Galadanci, N. A. (2020). Low educational level of head of household, as a proxy for poverty, is associated with severe anaemia among children with sickle cell disease living in a low-resource setting: evidence from the SPRING trial. *British Journal of Haematology*, *190*(6). 939-944. https://doi.org/10.1111/bjh.16746

Campbell, A., Rizio, A., McCausland, K., Iorga, S., Yen, G., Paulose, J., & Lee, S.(2023). The Burden of Sickle Cell Disease on Children and Their Caregivers: Caregiver Reports of Children's Health-Related Quality of Life and School Experiences, Caregiver Burden,

and Their Association with Frequency of Vaso-Occlusive Crises. *Patient Related Outcome Measures*, *Volume 14*. https://doi.org/10.2147/prom.s419607

Collier, R. (2012). The downside of genetic screening. 184(8), 862-864. https://doi.org/10.1503/cmaj.109-4169

Efe, I. (2013). Health care problem and management in Nigeria. *Journal of Geography and Regional Planning*, *6*(6), 244-254. https://doi.org/10.5897/jgrp2013.0366

Erusiafe, J., & Awunor, N. S. (2023). Knowledge, attitude and control practices of sickle cell disease among adults in rural communities in Udu LGA Delta State, Nigeria. *Annals of Clinical Sciences*, *8*(2), 58–67. https://acsjournal.lasucom.edu.ng/index.php/acs/article/view/153

Ezenwosu, O., Chukwu, B., Ezenwosu, I., Uwaezuoke, N., Eke, C., Udorah, M., Idoko, C., Ikefuna, A., & Emodi, I. (2021). Clinical depression in children and adolescents with sickle cell anaemia: influencing factors in a resource-limited setting. *BMC Pediatrics*, *21*(1). https://doi.org/10.1186/s12887-021-03015-1 https://doi.org/10.1186/s12887-021-03015-1

Feinstein, A. R. (1987). Clinimetric perspectives. *Journal of Chronic Diseases*, *40*(6). https://doi.org/10.1016/0021-9681(87)90027-0

Galadanci, N., Wudil, B. J., Balogun, T. M., Ogunrinde, G. O., Akinsulie, A., Hasan-hanga, F., Mohammed, A. S., Kehinde, M. O., Olaniyi, J. A., Diaku-akinwumi, I. N., Brown, B. J., Adeleke, S., Nnodu, O. E., Emodi, I., Ahmed, S., Osegbue, A. O., Akinola, N., Opara, H. I. O., Adegoke, S. A., …Adekile, A. D. (2014). Current sickle cell disease management practices in Nigeria. *International Health*, *6*(1). https://doi.org/10.1093/inthealth/iht022

Ibemere, S., Onyeka, T., Ezenwosu, O., Okoye, H. C., Ugwu, N., Tanabe, P. J., Shah, N., & Ezeanolue, E. (2023). Sickle Cell Disease Management Practices across Nigeria: A Cross-Sectional Analysis. *Blood*, *142*(Supplement 1). https://doi.org/10.1182/blood-2023-178129

Isa, H., Okocha, E., Adegoke, S. A., Nnebe-Agumadu, U., Kuliya-Gwarzo, A., Sopekan, A., Ofakunrin, A. O., Ugwu, N., Hassan, A. A., Ohiaeri, C., Madu, A., Diaku-Akinwumi, I., Ekwem, L., Dogara, L. G., Okoh, D., Jasini, J., Girei, A., Ekwere, T., Okolo, A., … Nnodu, O. (2023). Strategies to improve healthcare services for patients with sickle cell

disease in Nigeria: The perspectives of stakeholders. *Frontiers in Genetics*, *14*. https://doi.org/10.3389/fgene.2023.1052444

Kolapo, K.O, Vento, S. (2011). Stroke: a realistic approach to a growing problem in sub-Saharan Africa is urgently needed. *Trop Med Int Health*. 16(6):707-10. https://doi.org/10.1111/j.1365-3156.2011.02759.x

Leger, R. R., Wagner, L. D., & Odesina, V. (2018). Stigma in adults with sickle cell disease and family members: Scale development and pilot study in the USA and Nigeria. *International Journal of Africa Nursing Sciences*, *9*:23-29. https://doi.org/10.1016/j.ijans.2018.06.003

Lubeck, D., Agodoa, I., Bhakta, N., Danese, M., Pappu, K., Howard, R., Gleason, M., Halperin, M., Lanzkron, S. (2019). Life Expectancy and Income of Patients with Sickle Cell Disease Compared with Those Without Sickle Cell Disease Estimated.: *JAMA Netw Open.* 2(11):1915374. https://doi.org/10.1001/jamanetworkopen.2019.15374

Mahran, M. A., Ismail, M. T., & Abdelkader, E. H. (2019). 100 years of sickle cell disease research: etiology, pathophysiology and rational drug design (part 1). *Beni-Suef Univ J Basic Appl Sci* **8**, 22. https://doi.org/10.1186/s43088-019-0016-x

Masuwai, A., Zulkifli, H., & Hamzah, M. I. (2024). Evaluation of content validity and face validity of secondary school Islamic education teacher self-assessment instrument. *Cogent Education*, *11*(1). https://doi.org/10.1080/2331186X.2024.2308410

Makani, J., Soka, D., Rwezaula, S., Krag, M., Mghamba, J., Ramaiya, K., Cox, S. E., & Grosse, S. D. (2015). Health policy for sickle cell disease in Africa: Experience from Tanzania on interventions to reduce under-five mortality. *Tropical Medicine and International Health*, *20*(2):184-7. https://doi.org/10.1111/tmi.12428

Makani, J., Ofori-Acquah, S. F., Nnodu, O., Wonkam, A., & Ohene-Frempong, K. (2013). Sickle cell disease: New opportunities and challenges in Africa. *The Scientific World Journal*, *2013*. https://doi.org/10.1155/2013/193252

Moussa, M., Salifou, A. I., Nicolas, M., Pauline, Y., & Jean, K. (2023). Factors Associated with Non-Adherence to Treatment in Sickle Cell Patients Monitored at the National Reference Center for Sickle Cell Disease in Niger. *International Journal of Clinical Medicine*, *14*(02). https://doi.org/10.4236/ijcm.2023.142007

Modell, B., & Darlison, M. (2008). Global epidemiology of haemoglobin disorders and derived service indicators. *Bull World Health Organ* 86(6), 480-487. https://doi.org/10.2471/blt.06.036673

Mulumba, L.L., Wilson, L. (2015). Sickle cell disease among children in Africa: An integrative Literature review and global recommendations. *Internal Journal of African Nursing sciences*. 3, 56-54. https://doi.org/10.1016/j.ijans.2015.08.002

Nwagha, T., Omotowo, B.I.(2020). Determinants of Psychosocial Health-related Quality of Life of Adults with Sickle Cell Disease in a Nigerian Setting. *Niger Med J.*61(3): 114-119. https://doi.org/10.4103/nmj.NMJ_122_19

Ochei, M. (2024). WHO lauds Delta on universal health coverage. *Punch Newspaper [online]*. https://punchng.com/who-lauds-delta-on-universal-health-coverage/

Odame, I. (2010). Developing a Global Agenda for Sickle cell Disease: Report of an international symposium and Workshop in Cotonou, Republic of Benin. *American Journal of Preventive Medicine.* 38: S571-S575. https://doi.org/10.1016/j.amepre.2009.12.021

Odunvbun, M. E., Okolo, A. A., & Rahimy, C. M. (2008). Newborn screening for sickle cell disease in a Nigerian hospital. *Public Health*, *122*(10):1111-6. https://doi.org/10.1016/j.puhe.2008.01.008

Ogunseyin, O (2023). Delta State: Okowa signs nine bills into law. *The Guardian Nigerian News*. https://guardian.ng/news/delta-state-okowa-signs-nine-bills-into-law/

Okocha, E. C., Gyamfi, J., Ryan, N., Babalola, O., Etuk, E. A., Chianumba, R., Nwegbu, M., Isa, H., Madu, A. J., Adegoke, S., Nnebe-Agumandu, U., Brown, B., Peprah, E., & Nnodu, O. E. (2022). Barriers to Therapeutic Use of Hydroxyurea for Sickle Cell Disease in Nigeria: A Cross-Sectional Survey. *Frontiers in Genetics*, *12*, 765958. https://doi.org/10.3389/fgene.2021.765958

Olatunya, O. S., Babatola, A. O., Adeniyi, A. T., Lawal, O. A., Daramola, A. O., Agbesanwa, T. A., Ojo, T. O., Ajayi, P. O., Ibijola, A. A., Komolafe, A. K., & Adekile, A. (2021). Determinants of care-seeking practices for children with sickle cell disease in Ekiti, Southwest Nigeria. *Journal of Blood Medicine*, *12*. https://doi.org/10.2147/JBM.S294952

Oron, A. P., Chao, D. L., Ezeanolue, E. E., Ezenwa, L. N., Piel, F. B., Ojogun, O. T., Uyoga, S., Williams, T. N., & Nnodu, O. E. (2020). Caring for Africa's sickle cell children:

Will we rise to the challenge? *BMC Medicine*, *18*(1):92. https://doi.org/10.1186/s12916-020-01557-2.

Piel, F. B., Hay, S. I., Gupta, S., Weatherall, D. J., & Williams, T. N. (2013). Global Burden of Sickle Cell Anaemia in Children under Five, 2010-2050: Modelling Based on Demographics, Excess Mortality, and Interventions. *PLoS Medicine*, *10*(7). https://doi.org/10.1371/journal.pmed.1001484

Rodenberg CA (2009). A Review of: "Health Measurement Scales: A Practical Guide to Their Development and Use , Fourth Edition, by D. L. Streiner and G. R. Norman." *Journal of Biopharmaceutical Statistics*, *19*(6). https://doi.org/10.1080/10543400903244262

Salih, K.M.A. (2019). The impact of sickle cell anemia on the quality of life of sicklers at school age. *J Family Med Prim Care*. 8(2), 468-471. https://doi.org/10.4103/jfmpc.jfmpc_444_18.

Serjeant, G.R. (2010). One hundred years of sickle cell disease. Br J Haematol 151(5), 425-9. https://doi.org/10.1111/j.1365-2141.2010.08419.x

Tusuubira, S. K., Nakayinga, R., Mwambi, B., Odda, J., Kiconco, S., & Komuhangi, A. (2018). Knowledge, perception and practices towards sickle cell disease: A community survey among adults in Lubaga division, Kampala Uganda. *BMC Public Health*, *18*(1). https://doi.org/10.1186/s12889-018-5496-4

Uchechukwu, D. N. M. (2016). Knowledge of Sickle Cell Disease in a Rural Community of Delta State, South-South Nigeria. *IOSR Journal of Dental and Medical Sciences*, *15*(07). https://doi.org/10.9790/0853-15077100104

Vaske, J. J., Beaman, J., & Sponarski, C. C. (2016). Rethinking internal consistency in Cronbach's Alpha. *Leisure Sciences*. 39(2), 163-173. http://dx.doi.org/10.1080/01490400.2015.1127189

Zaki, R., Bulgiba, A., Ismail, R., & Ismail, N. A. (2012). Statistical methods used to test for agreement of medical instruments measuring continuous variables in method comparison studies: A systematic review. *PLoS One 7(5), e37908*. https://doi.org/10.1371/journal.pone.0037908.