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Clinical Profile, Management Patterns, and Outcomes of Sickle Cell Disease at a Tertiary Hospital in Nigeria

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Abstract Sickle cell disease (SCD) is a major public health concern in Nigeria, with a high prevalence and significant morbidity and mortality. Despite advances in management strategies, gaps in comprehensive care persist, particularly in resource-limited settings. This study aims to assess the clinical profile, management patterns, and outcomes of SCD patients at a tertiary hospital in Nigeria. A retrospective hospital-based audit was conducted at the University of Medical Sciences Teaching Hospital Complex, Ondo, Nigeria. Medical records of patients diagnosed with SCD were reviewed. Data collected included demographic characteristics, clinical presentations, frequency and types of crises, management strategies, and treatment outcomes. Descriptive and inferential statistical analyses were performed to identify predictors of adverse outcomes. A total of 71 patients were included in the study, with a mean age of 6.8 ± 3.8 years. Vaso-occlusive crises were the most common complication occurring in 74.6% of patients, followed by hemolytic (14.1%), sequestration (4.2%), aplastic (1.4%), and megaloblastic (2.8%) crises. None of the patients received hydroxyurea therapy, while folic acid (97.2%) and antimalarial prophylaxis (95.8%) were the most used medications. The mean number of blood transfusions per year was 1.2 (IQR: 1.0–2.0), and specialist consultations were infrequent. Logistic regression analysis identified lower lowest recorded packed cell volume (PCV) as a significant predictor of adverse outcomes (OR: 0.488, 95% CI: 0.286–0.835, p = 0.009). The findings highlight critical gaps in SCD management, including the underutilization of hydroxyurea and limited access to specialist care. Addressing these gaps through policy initiatives, provider education, and healthcare system strengthening could improve patient outcomes. These results provide valuable insights for optimizing SCD management in Nigeria.

Keywords: *sickle cell disease, vaso-occlusive crisis, management, Nigeria, hydroxyurea, blood transfusion, outcomes*

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1. Introduction

Sickle cell disease (SCD) is a hereditary hemoglobinopathy characterized by the production of abnormal hemoglobin S, leading to the deformation of erythrocytes into a sickle shape. These deformed red blood cells have reduced deformability, leading to recurrent vaso-occlusive crises (VOC), chronic hemolysis, and multi-organ complications [1]. The disease is inherited in an autosomal recessive manner, primarily affecting individuals of African, Mediterranean, Middle Eastern, and Indian ancestry [1]. It is estimated that approximately 300,000 infants are born with SCD annually worldwide, with sub-Saharan Africa bearing nearly 75% of this global burden [2].

Nigeria has the highest prevalence of SCD globally, with an estimated 2–3% of the population affected and about 20–30% carrying the sickle cell trait [3]. The high prevalence is largely due to the protective advantage conferred by the sickle cell trait against Plasmodium *falciparum* malaria, which is endemic in the region [3]. Despite increased awareness and newborn screening initiatives in some urban centers, SCD remains a significant cause of childhood morbidity and mortality in Nigeria. Studies have shown that without appropriate interventions, 50–80% of children born with SCD in sub-Saharan Africa do not survive beyond their fifth birthday [2]. These deaths are largely attributable to severe anemia, infections, and complications such as acute chest syndrome and stroke, which often occur due to inadequate healthcare infrastructure and limited access to comprehensive care [3].

While significant advancements have been made in the management of SCD, particularly in high-income countries, treatment options remain limited in resource-constrained settings. Hydroxyurea, an oral medication that increases fetal hemoglobin levels and reduces the frequency of VOC and hospitalizations, has been a major breakthrough in SCD management [4]. However, its accessibility in Nigeria is limited due to cost, lack of

physician awareness, and patient reluctance due to misconceptions about side effects [3]. Furthermore, curative treatment modalities such as hematopoietic stem cell transplantation (HSCT) and gene therapy remain largely inaccessible to most patients in sub-Saharan Africa due to high costs and infrastructure limitations [3].

In December 2023, the U.S. Food and Drug Administration (FDA) approved two revolutionary gene therapies, Casgevy (exagamglogene autotemcel) and Lyfgenia (lovotibeglogene autotemcel), which offer curative potential for patients with SCD [5]. These therapies involve genetic modification of a patient's hematopoietic stem cells to enable the production of functional hemoglobin, thereby reducing or eliminating disease manifestations. While these advances mark a transformative shift in the management of SCD, their applicability in Nigeria is highly limited by cost, lack of expertise, and logistical challenges. Consequently, the need for comprehensive audits of existing SCD cases within local healthcare settings is imperative to inform context-specific interventions.

This study aims to provide an audit of SCD cases seen at the University of Medical Sciences Teaching Hospital Complex, Ondo, Nigeria. The objectives are to evaluate patient demographics, clinical presentations, common complications, management strategies, and treatment outcomes. By identifying gaps in care and opportunities for improvement, this study seeks to provide valuable insights for optimizing SCD management protocols in Nigeria. Given the ongoing efforts to improve SCD care through policy interventions and the expansion of comprehensive care programs, findings from this study may contribute to the development of targeted strategies to enhance patient outcomes and quality of life in affected individuals.

2. Methods

2.1. Study Design

This study was a retrospective hospital-based audit conducted at the University of Medical Sciences Teaching Hospital Complex, Ondo, Nigeria. The study reviewed medical records of patients diagnosed with SCD over a specified period. Data were collected on patient demographics, clinical presentations, frequency and types of crises, management strategies, treatment interventions, and outcomes. The retrospective design allowed for an in-depth evaluation of the hospital's clinical and management trends, helping to identify gaps in care and potential areas for for improvement.

2.2. Study Setting

The study was conducted at the University of Medical Sciences Teaching Hospital Complex in Ondo, a tertiary-level healthcare institution that serves as a referral center for patients with various medical conditions, including hematological disorders. The hospital is one of the major centers in southwestern Nigeria that provides specialized care for individuals with SCD, offering outpatient follow-up services, emergency care for acute sickle cell crises, and inpatient management for complications associated with the disease. Additionally, the hospital provides supportive services such as blood transfusion, laboratory investigations, and consultations with specialists in cardiology, nephrology, and orthopedic surgery. The hospital's SCD clinic plays a critical role in the multidisciplinary management of patients by offering routine monitoring, early complication detection, and the administration of disease-modifying therapies such as hydroxyurea. Given the significant burden of SCD in the region, this healthcare facility was deemed an appropriate setting for conducting the study.

2.3. Study Population and Eligibility Criteria

The study population consisted of all patients diagnosed with SCD who received medical care at the University of Medical Sciences Teaching Hospital Complex during the study period. Only patients with a confirmed diagnosis of SCD were included, with confirmation based on laboratory findings from hemoglobin electrophoresis or high-performance liquid chromatography (HPLC). To ensure data reliability, only cases with complete medical records, including demographic details, clinical history, and treatment documentation, were considered eligible for inclusion.

Patients were included in the study if they had at least one documented clinical visit during the study period and met the diagnostic criteria for SCD. Cases were excluded if records were incomplete or if the diagnosis was ambiguous due to coexisting hemoglobinopathies such as β-thalassemia major without concurrent SCD. The inclusion of only well-documented cases ensured that the data analysis was based on high-quality, verifiable clinical information, thereby enhancing the study's validity and reliability.

2.4. Sample Size Determination

The sample size for this study was determined using Cochran's formula for sample size estimation in cross-sectional studies. Given that the prevalence of SCD in Nigeria has been estimated to be approximately 2.5% [3], this value was used as the basis for sample size estimation. A 95% confidence level was chosen to ensure statistical reliability, corresponding to a standard normal deviate of 1.96 [6]. The margin of error was set at 5% to balance precision and feasibility. Using these parameters, the initial sample size calculation suggested that at least 38 patients would be required to achieve a statistically robust analysis.

However, since this was a hospital-based study with a finite population of patients receiving care at the University of Medical Sciences Teaching Hospital Complex, an adjustment using the finite population correction formula was applied. The hospital records indicated that approximately 250 to 300 patients with SCD were managed during the study period. Incorporating this information into the sample size adjustment reduced the estimated minimum required sample size to 33. To ensure robustness, accommodate potential missing or incomplete data, and enhance the generalizability of findings, the final sample size was rounded up to a minimum of 40 patients.

However, given the retrospective nature of the study, all available and eligible cases within the study period were included in the analysis to maximize data utilization. This approach strengthened the validity of the findings by increasing statistical power and reducing the risk of selection bias.

2.5. Data Collection and Variables

Data were extracted from patient medical records and the hospital's electronic health system using a structured data collection form. The information collected included demographic characteristics such as age, sex, genotype classification (HbSS, HbSC, or HbSβ-thalassemia), and parental educational background. Clinical data were recorded, including the age at diagnosis, history of sickle cell crises, and frequency of hospital admissions. The specific types of crises experienced by patients, including vaso-occlusive crises, hemolytic crises, sequestration crises, aplastic crises, and megaloblastic crises, were documented.

Management data included the routine use of medications such as hydroxyurea, folic acid, multivitamins, and antimalarial prophylaxis. The frequency of blood transfusions was recorded, along with details of any specialist consultations, such as ophthalmology evaluations for retinopathy, cardiology assessments for cardiac complications, and orthopedic interventions for avascular necrosis. Laboratory parameters, including packed cell volume (PCV) at steady state and during crises, full blood count (FBC), transcranial Doppler (TCD) screening, echocardiography, and imaging studies such as CT scans and chest X-rays, were also extracted.

The primary outcomes assessed included the stability of patients on follow-up, the presence of complications, loss to follow-up, and mortality. Outcomes were categorized based on whether patients remained clinically stable, developed complications requiring further intervention, or were lost to follow-up. Mortality data were reviewed to assess the leading causes of death in the studied population.

2.6. Data Analysis

Data analysis was conducted using IBM SPSS Statistics (version 21.0, IBM Corp, Armonk, NY, USA). Descriptive statistics were used to summarize patient demographics, clinical presentations, management patterns, and outcomes. Categorical variables were expressed as frequencies and percentages, while continuous variables were assessed for normality and summarized using means with standard deviations for normally distributed data or medians with interquartile ranges for skewed distributions.

Comparative analyses were performed to assess differences in clinical outcomes based on patient demographics and disease severity. The chi-square test or Fisher's exact test was used to compare categorical variables, while independent t-tests or Mann-Whitney U tests were applied for continuous variables depending on their distribution. Logistic regression analysis was conducted to determine potential predictors of adverse outcomes, including frequent hospitalizations, severe complications, and mortality. The significance threshold was set at a p-value of less than 0.05 for all statistical tests.

2.7. Ethical Considerations

Ethical approval for the study was obtained from the Ethics Review Committee of the University of Medical Sciences Teaching Hospital Complex, Ondo. Given the retrospective nature of the study, informed consent was not required from patients. However, patient confidentiality was strictly maintained by anonymizing all identifying information during data collection and analysis. The study adhered to ethical principles outlined in the Declaration of Helsinki for research involving human subjects, ensuring that the rights and privacy of patients were respected throughout the research process.

3. Results

3.1. Demographic and Clinical Characteristics of the Study Population

A total of 71 patients were included in the study. The mean age of the study population was 6.8 ± 3.8 years, while the median age was 6.0 years with an interquartile range of 4.0 to 9.25 years. The majority of the patients were male, accounting for 60.6% (n = 43), while females comprised 38.0% (n = 27) (Table 1).

Regarding hemoglobin genotype distribution, 93.0% (n = 66) of the patients had HbSS, while 4.2% (n = 3) had HbSC and 2.8% (n = 2) had HbSβ-thalassemia. The mean age at diagnosis of SCD was 3.3 ± 2.4 years. Assessment of parental educational background showed that 16.9% (n = 12) of the mothers had tertiary education, 5.6% (n = 4) had secondary education, and none had only primary education. Among fathers, 7.0% (n = 5) had tertiary education, 4.2% (n = 3) had secondary education, and none had only primary education (Table 1).

Table 1. Demographic and Clinical Characteristics of the Study Population

Variable	Value
Total Patients	71
Age (Mean ± SD)	6.8 ± 3.8 years
Age (Median, IQR)	6.0 (4.0 - 9.25) years
Male, n (%)	43 (60.6%)
Female, n (%)	27 (38.0%)
Hemoglobin types	
HbSS, n (%)	66 (93.0%)
HbSC, n (%)	3 (4.2%)
HbSβ-thal, n (%)	2 (2.8%)
Age at Diagnosis (Mean ± SD)	3.3 ± 2.4 years
Mother's Education, n (%)	
Tertiary	12 (16.9%)
Secondary	4 (5.6%)
Primary	0 (0.0%)
Father's Education, n (%)	
Tertiary	5 (7.0%)
Secondary	3 (4.2%)
Primary	0 (0.0%)

3.2. Frequency and Types of Sickle Cell Crises

The frequency and types of sickle cell crises experienced by the study population were assessed. The mean number of crises per year among patients was 1.3, with an interquartile range of 1.0 to 1.0. The mean number of hospital admissions due to crises per year was 1.1, with an interquartile range of 1.0 to 1.0 (Table 2).

Vaso-occlusive crises were the most frequently reported type, occurring in 74.6% (n = 53) of patients. Hemolytic crises were documented in 14.1% (n = 10) of cases. Sequestration crises, aplastic crises, and megaloblastic crises were also recorded among the study population, with their respective frequencies detailed in Table 2.

Table 2. Frequency and Types of Sickle Cell Crises Among Patients

Variable	Value
Total Patients	71
Mean Crises per Year (Mean, IQR)	1.3 (1.0 - 1.0)
Mean Hospital Admissions per Year (Mean, IQR)	1.1 (1.0 - 1.0)
Vaso-Occlusive Crisis, n (%)	53 (74.6)
Hemolytic Crisis, n (%)	10 (14.1)
Sequestration Crisis, n (%)	3 (4.2)
Aplastic Crisis, n (%)	1 (1.4)
Megaloblastic Crisis, n (%)	2 (2.8)

3.3. Treatment and Management Strategies

The use of routine medications among the study population varied. Folic acid was the most commonly used medication, with 97.2% (n = 69) of patients receiving it. Antimalarial prophylaxis was documented in 95.8% (n = 68) of patients. None of the patients were recorded as using hydroxyurea or multivitamins, while Ciklavit was used by 1.4% (n = 1) of patients (Table 3).

Table 3. Treatment and Management Strategies Among Patients with Sickle Cell Disease

Variable	Value
Total Patients	71
Folic Acid Use, n (%)	69 (97.2)
Multivitamin Use, n (%)	0 (0.0)
Antimalarial Use, n (%)	68 (95.8)
Hydroxyurea Use, n (%)	0 (0.0)
Ciklavit Use, n (%)	0 (0.0)
Mean Blood Transfusions per Year (Mean, IQR)	1.1 (1.0 - 1.0)
Ophthalmologist Consultation, n (%)	0 (0.0)
Cardiologist Consultation, n (%)	1 (1.4)
Orthopedic Consultation, n (%)	1 (1.4)
ENT Consultation, n (%)	1 (1.4)

The mean number of blood transfusions per year among patients was 1.2, with an interquartile range of 1.0 to 2.0. Laboratory and imaging investigations performed included transcranial Doppler screening, echocardiography, CT scans, full blood count analysis, and chest X-rays, with details presented in Table 3.

Specialist consultations were also assessed. Ophthalmologist consultations were recorded in 5.6% (n = 4) of patients, while 4.2% (n = 3) had a cardiology consultation. Orthopedic consultations were documented in 1.4% (n = 1), and ENT consultations in 1.4% (n = 1) of the patients (Table 3). As at the time of this study one paitent had died of cerebrovascular accident.

3.4. Laboratory Findings and Disease Severity

The steady-state packed cell volume (PCV) of the study population had a mean value of 24.5%, with an interquartile range of 21.0% to 28.0%. The lowest recorded PCV had a mean value of 17.5%, with an interquartile range of 13.0% to 22.0% (Table 4). The mean number of blood transfusions per year among patients was 1.1. (Table 4).

Complications were observed in 16.9% (n = 12) of the study population. Stroke was recorded in 2.8% (n = 2) of patients, while acute chest syndrome was reported in 4.2% (n = 3). Avascular necrosis was documented in 1.4% (n = 1) of patients. Other complications were present in 8.5% (n = 6) (Table 4).

Table 4. Laboratory Findings and Disease Severity Indicators

Variable	Value
Total Patients	71
Steady-State PCV (Mean, IQR)	24.5 (21.0 - 28.0)
Lowest Recorded PCV (Mean, IQR)	17.5 (13.0 - 22.0)
Mean Blood Transfusions per Year (Mean, IQR)	1.1 (1.0 - 1.1)
Patients with Complications, n (%)	12 (16.9)
Stroke, n (%)	2 (2.8)
Acute Chest Syndrome, n (%)	3 (4.2)
Avascular Necrosis, n (%)	1 (1.4)
Other Complications, n (%)	6 (8.5)

3.5. Predictors of Adverse Outcomes

The relationship between demographic and clinical factors and the occurrence of adverse outcomes was examined. Adverse outcomes were defined as the need for at least one blood transfusion and/or a specialist consultation. Logistic regression analysis was performed to determine the predictors of these adverse outcomes (Table 5).

Table 5. Predictors of Adverse Outcome among Patients

Variable	Odds Ratio	95% CI Lower	95% CI Upper	p
Age (years)	0.996	0.746	1.328	0.977
Number of Crises per Year	0.759	0.193	2.985	0.693
Stable-State PCV (%)	0.935	0.701	1.245	0.644
Lowest Recorded PCV (%)	0.488	0.286	0.835	0.009

The odds of experiencing an adverse outcome were lower with Increasing lowest recorded packed cell volume (PCV), with an odds ratio of 0.488 (95% CI: 0.286 – 0.835, p = 0.009). The odds of an adverse outcome were also examined in relation to age, number of crises per year, and stable-state PCV, but no statistically significant associations were observed (Table 5).

These findings summarize the clinical and demographic factors associated with adverse outcomes in the study population. Further details on the logistic regression analysis and statistical comparisons are presented in Table 5.

4. Discussion

The findings from this study revealed that VOCs were the predominant complication, occurring in 74.6% of patients. This aligns with existing literature, as VOCs are a hallmark of SCD, resulting from microvascular obstruction and ischemia-induced pain episodes [1]. Other crises, such as hemolytic, sequestration, aplastic, and megaloblastic, were observed less frequently, reflecting the diverse clinical spectrum of SCD.

Notably, none of the patients in this cohort were receiving hydroxyurea therapy, despite its proven efficacy in reducing VOC frequency and transfusion requirements [7]. This underutilization may be attributed to factors such as cost, limited awareness among healthcare providers and patients, and concerns about potential adverse effects [4]. In contrast, folic acid supplementation and antimalarial prophylaxis were widely prescribed, consistent with standard care practices in malaria-endemic regions to prevent folate deficiency and malaria-related complications [8].

Blood transfusions were required in 29.6% of patients, with an average of 1.1 transfusions per year. While transfusions are a critical component of SCD management, especially in severe anemia or acute complications, the relatively low transfusion rate observed may reflect either a less severe disease phenotype in this cohort or potential underutilization due to resource constraints [9].

Specialist consultations were notably scarce; only 4.2% of patients were referred to a cardiologist, and even fewer accessed ophthalmology, orthopedic, or ENT services. This indicates a potential gap in comprehensive SCD care, as multidisciplinary management is essential for early detection and treatment of complications such as retinopathy, pulmonary hypertension, and avascular necrosis [10]. Barriers to specialist access may include systemic healthcare limitations, financial constraints, or lack of structured referral pathways.

Logistic regression analysis identified a lower lowest recorded PCV as a significant predictor of adverse outcomes, with an odds ratio of 0.488. This finding is consistent with prior studies demonstrating that severe anemia correlates with increased morbidity and the need for interventions such as transfusions [11]. Other factors, including age, number of crises per year, and stable-state PCV, were not significantly associated with adverse outcomes in this cohort.

These results underscore the need for improved SCD management strategies in Nigeria. The absence of hydroxyurea use suggests a critical area for intervention, including provider education, cost reduction initiatives, and community awareness programs to increase uptake [12]. Enhancing access to multidisciplinary care through better referral systems and healthcare infrastructure could facilitate early complication detection and management, potentially improving patient outcomes [13]. Additionally, the association between low PCV and adverse outcomes highlights the importance of regular hematologic monitoring and timely interventions to mitigate risks.

This study has limitations, including its retrospective design and single-center setting, which may limit the generalizability of the findings. Incomplete medical records could introduce bias, and the sample size, while sufficient for analysis, may not capture the full spectrum of SCD complications across different healthcare settings. Future research should consider larger, multicenter studies to enhance external validity and explore barriers to hydroxyurea use among both healthcare providers and patients. Investigating the feasibility of structured comprehensive care models, including routine specialist consultations, is also warranted. Prospective cohort studies examining the long-term impact of transfusions, hydroxyurea, and multidisciplinary interventions on patient outcomes would provide valuable insights. Assessing the role of emerging gene therapies in the Nigerian context could help shape future treatment strategies [14].

In conclusion, this study provides valuable insights into the clinical profile, management patterns, and outcomes of SCD patients in Nigeria. The findings highlight critical areas for improvement, particularly in the uptake of hydroxyurea, access to specialist care, and transfusion practices. Addressing these gaps through policy initiatives, provider education, and improved healthcare infrastructure could significantly enhance SCD care and patient outcomes in Nigeria.

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