Review Article *Open Access*

Journal of Innovative Clinical Trials and Review Articles

ISSN: 3068 - 1936

DOI: doi.org/10.63721/26JCTC0155

Burden of Sickle Cell Disease in Paediatric Patients Admitted at Rundu Intermediate State Hospital Over a 2-Year Period

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Citation: *Clothilde Natasha Sanzila, Runyararo Mashingaidze-Mano (2026) Burden of Sickle Cell Disease in Paediatric Patients Admitted at Rundu Intermediate State Hospital Over a 2-Year Period. J. of Inn Clin Trail Case Reports 2(5), 1-7. WMJ/JCTC-155*

Abstract

Background: *The origin of sickle cell disease (SCD) lies in the malarial regions of the tropics, where carriers are protected against death from malaria and hence enjoy an evolutionary advantage. The βS-mutation is the archetypal example of natural selection in humans. Of the total population of Namibia (2.46 million), 72% are living in areas of active malaria transmission. Patients with SCD suffer from various clinical manifestations.*

Methods: *This was a retrospective cross-sectional study using paediatric hospital records for patients admitted to the Rundu State Intermediate Hospital in 2020 and 2021.*

Results: *Over the course of two years (2020–2021), a total number of 3,168 children were admitted to the Rundu state hospital, with 1,527 and 1,641 admitted in 2020 and 2021, respectively. Of the total admissions, 103 of the paediatric patients had SCD, which constituted a prevalence of 3.3%. Many of the participants were male, aged between 1 and 5 years old, and suffering mainly from vaso-occlusive crisis.*

Conclusion: *The burden of SCD among admitted patients at Rundu hospital was high, and vaso-occlusive crisis was the main clinical manifestation. Patients with SCD had access to adequate management. There is a need to diagnose SCD in newborns before the development of complications, and comprehensive care must be offered in the future in Namibia.*

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Submitted: 17.09.2026 **Accepted**: 21.09.2026 **Published**: 30.09.2026

Keywords: Sickle Cell Disease, Burden, Children, Admitted, Namibia

Introduction

Sickle cell disease (SCD) is an autosomal recessive multisystem disorder characterized by chronic haemolytic anemia, painful ischemic episodes of vaso-occlusion, and progressive organ failure [1]. The term "sickle cell disease" refers to any condition in which the production of sickle haemoglobin (HbS) leads to pathophysiological consequences [2]. Sickle haemoglobin is a structural variant of normal adult haemoglobin (HbA) caused by a mutation in the haemoglobin subunit beta (HbB) gene, which leads to the substitution of valine for glutamic acid at position 6 of the β-globin subunit (βS) of the haemoglobin molecule [3]. The frequency of this gene is highest in West African countries, with 1 in 3 or 4 (25–30%) people being carriers of HbS, compared to 1 in 400 African Americans and variable frequency in European populations [4]. According to Jastaniah [5], the prevalence of the sickle cell trait ranges between 10% and 40% across equatorial Africa and decreases to between 1% and 2% on the North African coast and less than 1% in South Africa. The vast majority of SCD births occur in sub-Saharan Africa [1]. The prevalence of SCD in developed countries is increasing partly due to migration from high-prevalence countries [5]. It is estimated that over 14,000 people live with SCD in the UK, similar to France, while countries like Italy and Germany have also seen increasing numbers of migrants from Africa [4].

Patients may be completely asymptomatic during the first 6 months of life due to the presence of fetal haemoglobin, which gradually decreases, and HbS begins to predominate [6]. Clinical manifestations are variable, affect multiple organ systems, and generally cause lower life expectancy [6]. Children with sickle cell disease can present with a range of clinical manifestations, including ocular findings such as retinal vascular tortuosity [7], and acute events such as painful crises, acute chest syndrome, and infections [8]. These children are also at risk of stroke and recurrent acute chest syndrome, which are the main clinical problems in severe cases [9]. Other common manifestations include musculoskeletal involvement, gastrointestinal symptoms, and genitourinary complications [10]. The management of these manifestations often involves a combination of therapies, including hydroxyurea, blood transfusion, and bone marrow transplantation [8].

Children with SCD are at a higher risk of developing severe complications from malaria, including fever, pallor, and jaundice [11]. Asymptomatic malaria infection is also prevalent in this population, potentially due to elevated levels of IL-10 [12]. However, the prevalence of malaria in children with SCD may not be significantly higher than in those without SCD, and the risk of severe malaria may be lower [13].

Research on severe bacterial infections in children with sickle cell disease has shown varying incidence rates and outcomes. A study by Bansil in 2013 found an overall incidence rate of 16.0%, with pneumonia being the most common infection. Another study Gbadoé in 2023 reported a decline in Salmonella and an increase in Escherichia coli, Klebsiella pneumoniae, and Staphylococcus aureus [14, 15].

In high-income countries, the increase in HbS largely reflects gains in life expectancy among affected persons as a result of interventions such as newborn screening, penicillin prophylaxis, primary stroke prevention, and hydroxyurea treatment [11]. Life expectancy has improved significantly in high-income countries over the past 40 years, with childhood mortality now close to that in the general population and an observed median survival of more than 60 years [11]. In Africa, where there is a lack of newborn screening and routine childhood vaccinations and where malaria, malnutrition, and poverty remain important challenges, the mortality among children with sickle cell disease who are younger than 5 years of age can be as high as 90% [11].

The origin of SCD lies in the malarial regions (of the tropics) in which carriers are protected against death from malaria and hence enjoy an evolutionary advantage [1]. The βS -mutation is the archetypal example of natural selection in humans [16]. Heterozygotes, whose red blood cells contain both HbA and HbS, are so strongly protected from malaria that the global distribution and the frequency of the βS -mutation a mutation now strongly reflects the historic incidence of death from malaria [2]. Uyoga et al [17], explained

that the distribution reflects the fact that sickle-cell trait confers a survival advantage against malaria and that selection pressure due to malaria has resulted in high frequencies of the mutant gene, especially in the areas of high malarial transmission. Although a single abnormal gene may protect against malaria, inheritance of two abnormal genes leads to SCD and confers no such protection, and malaria is one of the major causes of morbidity and death in children with SCD in Africa [5].

Seventy two percent (72%) of the total population of Namibia are living in areas of active transmission of malaria (Total population: 2.46 million) [18]. Most of Namibia's malaria cases occur during the rainy season between January and April, in the northern region of the country [18]. Although Namibia has made significant progress over the last decade, the ongoing threat of malaria importation from across the country's borders is a key barrier to achieving elimination [18].

As new therapies emerge, potentially leading to disease amelioration or cure, it is of paramount importance that the significant burden of SCD in resource-poor countries is properly recognized [2]. The environmental conditions that lead to emergence of the S mutation insure (HbS) – at least in equatorial Africa - that the highest prevalence of SCD is found in regions where the malaria parasite is still endemic [11].

Sickle cell disease (SCD) is an increasing global health problem [11]. It is a major, but widely neglected, public health issue in low-income countries [17], Namibia included. The prevalence of sickle cell disease in children in Namibia is unknown. The World Health Organization has declared Sickle Cell Anemia (SCA) a public health priority [19,20]. In Africa, 50–90% of children born with sickle cell disease die before they reach their fifth birthday [17], most of them present with different crisis. It is known that the area of high malarial burden is directly proportional to the prevalence of sickle cell disease. Rundu has high malaria [21]. There is lack of adequate data in Namibia on children affected with disease and even mortality, so this study is going to address the gap in knowledge.

The aim of the study was to document clinical presentation and treatment outcome of paediatric patients with sickle cell disease admitted at the Rundu intermediate hospital in 2020 and 2021.

Methodology

This was a retrospective cross-sectional study, which used hospital records of paediatric patients of 12 years and below admitted with a diagnosis of sickle cell disease at the Rundu Intermediate Hospital from 1 January 2020 to 31 December 2021. Data was collected from 1 August to 30 September 2022. Formal consent for the study was obtained from the Ministry of Health and Social Services (MOHSS)'s Biomedical Research Ethics Committee (BREC) and Research Management Committee (RMC) in the Office of the Executive Director. Of note, MOHSS's Biomedical Research Ethics Committee (BREC) is an entity under MOHSS. Consent was wavered as data was collected from the hospital records.

Data Collection and Analysis Procedure

Recording sheets were used to abstract the data from the hospital database for each participant The demographic data which includes age, sex and place of origin were collected as well as clinical presentation (crisis: vaso-oclussive, haemolytic, sequestration, aplastic); malaria illness, and serious infection. Additionally, information on the medication (hydroxyurea, oral penicillin) and treatment outcome (death /discharge) were captured. An excel spreadsheet was created, data anonymized and cleansed then transferred to SPSS version 26.0 for analysis.

Results

Demographic Characteristics of Study Participants

Over the course of two years (2020-2021) a total number of 3168 children were admitted at the Rundu state hospital. From the total admissions 103 of the paediatric patients had sickle cell disease which constituted a prevalence of 3.3%. The total population of children less than 15 years in the Kavango East region is 93 921, which accounts for 43% of the youth population. Among the patients with SCD 38 (36.9%) were females and 65 (63.1%) males. The age range was 1 to 12 years with a median age of 5 years. Majority of the children, 54% were 5 years and below, with the remaining 12% and 34%, 6 to 10 years, 11 to 15 years respectively. The area of residence was categorized according to if patient was from rural or urban Kavango East. The urban area had the majority of the participants, a total of (n=65),

minority at (n=33) from rural, with 5 participants do not have clear documentation of area of residence. All the participants were Namibians specifically the Kavango tribe but are fairly distributed within the region. See Table 1 below.

Table 1: Demographic characteristics of children admitted with SCD at Rundu Intermediate State Hospital

Variables	Percentage (%)
	N=103
Sex	
Female	36.9
Male	63.1
Age at presentation	
1-5 years	54.4
6-10 years	33.9
11-15 years	11.7
Area of Residence	
Urban	63.1
Rural	32
Missing	4.9

Clinical Presentation of Patients with Sickle Cell Disease

Most participants presented in a Vaso-occlusive crisis, 62.1% and haemolytic crisis, 27.2%. Three patients (2.9%) presented with both crises, Vaso-occlusion and haemolytic. Eight (7.8%) patients presented with other medical conditions namely epilepsy, stroke, cerebral palsy, gastroenteritis and pneumonia (Fig. 1).

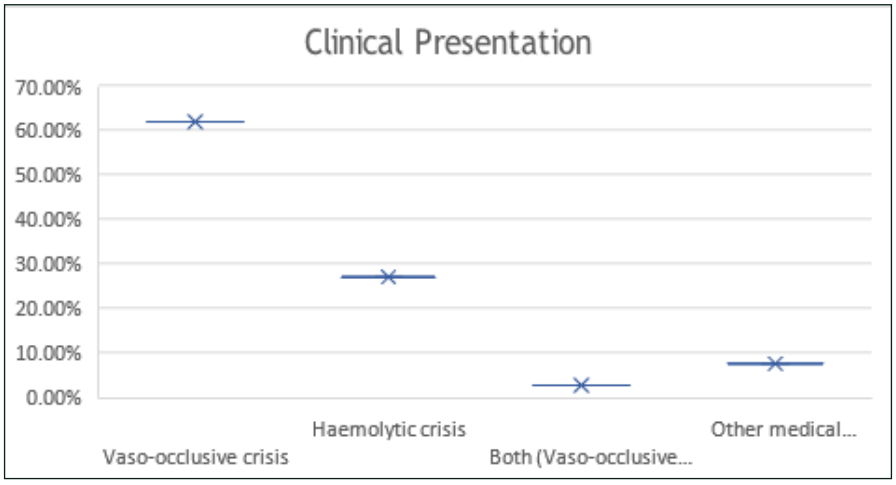

Figure 1: Crisis presentation for patients with sickle cell disease admitted at Rundu Intermediate State Hospital

One (1%) of participants had malaria, and 15 (14.6%) presented with an infection. Twenty-two (21.4%) patients had multiple admission whilst eighty-eight (78.6%) patients had only one admission Table 2 below. None of the participants died during the study period.

Table 2: Malaria, other infections and admissions rates of children with sickle cell disease

Variable	Percent (%)
	N=103

Malaria	
Yes	1
No	99
Infection	
Yes	14.6
No	85.4
Multiple admissions	
Yes	21.4
No	78.6

Majority of the participants 87.4%) received analgesics for pain, with 67.9% of them given opioid Antibiotics were administered to 96.1% received antibiotics, the majority received oral penicillin (pen VK) and others ceftriaxone. Twenty-eight patients (27.2%) required red blood transfusion. Patients who presented in vaso-oclussive crisis were given intravenous fluids. All patients had access to hydroxyurea.

Discussion

This study is the first to document clinical characteristics of patients with SCD in Namibia. The study showed the majority of the participants are boys 63.1% and 36.9% girls. These findings are similar to a study done by Mustafa et al [9] were 75 children among the study group, 34 (45%) were girls and 41 (55%) were boys. This may reflect SCD being common in boys than girls.In this study the majority of the children with SCD were 5 years and below. Similar findings were reported by Mustafa et al [9] with 52% of participants were 5 years and below. This may mean more children are diagnosed early as they suffer from different crises.

The majority of the participants came from urban 63.1% compared to 32% from rural and 5% with missing residence data respectively. This may mean those who are in urban area have easy access to the health facilities compared to the rural counterparts. Inequalities in income distribution, urban-rural disparities, and limited access to healthcare in remote areas are persistent (23, 24). These factors can delay diagnosis and treatment of individuals affected by sickle cell disease especially in regions where healthcare services are scarce.Most participants presented in a vaso-occlusive crisis, a total of 62.1% and haemolytic crisis of 27.2%. Painful vaso-occlusive crisis due to bony infarction is the commonest cause for hospital admissions in other studies [2]. Infectious diseases, such as malaria and pneumococcal disease (meningitis, pneumonia, and septicaemia) are thought to be the major cause of morbidity and mortality [1]. One (1%) of participants had malaria, and (n=15, 14.6%) presented with an infection. Despite Kavango region being considered a high malaria region, diagnosis of malaria in only 1% of the study participants may be a reflection in success in combating malaria in Namibia.

A study conducted at a university teaching hospital in Tanzania reported a SCD mortality rate of 1.9 per 100 person years of observation (PYO), with the highest incidence of death occurring in the first five years of life [16]. In low resource settings and countries where newborn screening is not yet standard care, patients may die young even before diagnosis is confirmed [4].Twenty-eight patients (27.2%) received red cell concentrate. Pain usually accounts for the majority of hospitalizations and overall negative impact in patients' health related quality of life [4]. Pain is the cardinal feature of SCD, and it is characteristically unpredictable, episodic in nature, described as one of the most excruciating forms of pain that affects human beings [4]. As described in results, opioids and other forms of analgesia were given to alleviate pain. Sickle cell disease Increases susceptibility to infections, notably bacterial sepsis and malaria in children under five years [4]. Respiratory infections can trigger the sickle-cell acute chest syndrome, with a high risk of death. Pain is the commonest acute complication of SCD and significantly impact health-related quality of life [4]. Patients with any form of infection had access to antibiotics. Additionally, essential medicine for SCD such as folic acid, oral penicillin (Pen VK) and hydroxyurea

were available to all patients. Hydroxycarbamide (or hydroxyurea) remains the only agent that has been proven to reduce the number of episodes of painful crises, acute chest syndrome and hospitalizations in randomized control trials in adults [2]. In addition, intravenous fluids were available for the participants. Given the fact that pain is often triggered by infection, exposure to cold or dehydration, supportive care during these episodes involves providing hydration, warmth and treating the underlying infection [4]. Currently the only available disease-modifying medications for SCD are hydroxycarbamide and l-glutamine [4]. Both are given daily to reduce the rate of acute complications, but results vary from person to person [4]. Another effective disease modifying therapy is blood transfusion to raise the haemoglobin for improved oxygenation in severe anaemia and also to reduce the proportion of sickle haemoglobin (HbS%), it may be given as a simple top-up blood transfusion or as exchange transfusion (manual or automated) [4]. The main curative therapy is stem cell transplantation while gene therapy has recently been approved by the United States Food and Drug Administration (FDA) [4, 22]. Despite the use of disease modifying therapy 21.4 % of participants had multiple admissions. Definitive treatment such as gene therapy and stem cell transplantation are areas for possible future expansion. Moreover, early diagnosis in newborn may help in intervening early before development of serious complications.

The limitation of the study was that it focused on the patients admitted at the Rundu Intermediate State Hospital in the Kavango East region, excluding patients seeking health services elsewhere including private sector, in addition, this was a retrospective study that used hospital files, some of which had missing data. To our knowledge this is the first study to document SCD burden in children in Namibia.

Conclusion

In conclusion SCD remains a public health problem affecting 3.3% of admission at Rundu Intermediate hospital. Children below the age of 5 years are mostly affected. Patients with SCD presented with various crisis and had access to adequate treatment in Namibia. Future studies should focus on creation of registries for paediatrics patients to enable proper follow up. There is need to improve record keeping of patient information to avoid gaps in future research. Additionally, consideration for prospective study exploring newborn screening in future is recommended.

Declarations

This study was carried out as partial fulfillment of the Bachelor of Medicine & Surgery at the University of Namibia

Acknowledgements

The contributions by the following individuals to this study are highly recognized, Dr Runyararo Mashingaidze Mano for supervising this study and providing guidance, Dr. Kafuro Adeline for assisting with data collection and Mr. Amukongo Mondjila for guidance with data analysis.

Authors' Contributions

CNS designed the study and drafted the paper in collaboration with RMM. CNS collected the data and then performed the analyses and interpreted the results. Both authors read and approved the final manuscript.

Funding

None

Conflict of interest

The authors declare that there was no conflict of interest.

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