Burden of sickle cell disease in paediatric patients admitted to Rundu Intermediate State Hospital, Namibia, over the 2-year period 2020 - 2021

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Background. Sickle cell disease (SCD) originated 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. Individuals with SCD suffer from various clinical manifestations.
Objectives. To document the clinical presentation and treatment outcome of paediatric patients with SCD admitted to Rundu State Intermediate Hospital in 2020 and 2021.
Methods. This was a retrospective cross-sectional study using paediatric hospital records for patients admitted to Rundu Hospital.
Results. Over the 2-year study period, a total of 3 168 children were admitted to Rundu Hospital, with 1 527 and 1 641 admitted in 2020 and 2021, respectively. Of these children, 103 had SCD, which constituted a prevalence of 3.3%. Many of the children with SCD were male, aged between 1 and 5 years, and suffering 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. SCD should be diagnosed in newborns before the development of complications, and comprehensive care must be offered in the future in Namibia.

Keywords. Sickle cell disease, burden, children, admitted, Namibia

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Sickle cell disease (SCD) is an autosomal recessive multisystem disorder characterised by chronic haemolytic anaemia, painful ischaemic 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 one in every three or four people (25 - 30%) being carriers of HbS, compared with 1 in 400 African Americans and variable frequency in European populations.[4] According to Jastaniah,[5] the prevalence of the sickle cell trait ranges from 10% to 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 owing 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 such as 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 owing to the presence of fetal haemoglobin. As this gradually decreases,

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HbS begins to predominate.[6] Clinical manifestations are variable, affect multiple organ systems, and generally cause reduced life expectancy.[6] Children with SCD 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 increased risk of developing severe complications from malaria, including fever, anaemia and jaundice.[11] Asymptomatic malaria infection is also prevalent in this population, potentially owing to elevated levels of interleukin-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 SCD has shown varying incidence rates and outcomes. A study by Bansil *et al.*[14] in 2013 found an overall incidence of 16.0%, with pneumonia being the most common infection. In 2023, Gbadoé *et al.*[15] reported a decline in *Salmonella* infections and an increase in *Escherichia coli*, *Klebsiella pneumoniae* and *Staphylococcus aureus*.

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 malaria, malnutrition and poverty remain important challenges, the mortality among children with SCD who are younger than 5 years of age can be as high as 90%.[11]

SCD originated 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 frequency of the βS mutation now strongly reflects the historical incidence of death from malaria.[2] Uyoga *et al.*[17] explained that the distribution reflects the fact that the sickle cell trait confers a survival advantage against malaria, and selection pressure due to malaria has resulted in high frequencies of the mutant gene, especially in 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 of the 2.46 million total population of Namibia live in areas of active transmission of malaria.[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 past 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 amelioration or cure of SCD, it is of paramount importance that the significant burden it imposes in resource-poor countries is recognised.[2] The environmental conditions that lead to emergence of the βS mutation – at least in equatorial Africa – ensure that the highest prevalence of SCD is found in regions where the malaria parasite is still endemic.[11]

SCD is an increasing global health problem.[11] It is a major but widely neglected public health issue in low-income countries,[17] including Namibia. The prevalence of SCD in children in Namibia is unknown. The World Health Organization has declared sickle cell anaemia a public health priority.[19,20] In Africa, 50 - 90% of children born with SCD die before they reach their 5th birthday,[17] presenting with a range of health crises. It is known that the burden of malaria is directly proportional to the prevalence of SCD. Rundu is a high-malaria area.[21] There is a lack of adequate data in Namibia on children with SCD, and even on mortality, so this study at Rundu State Intermediate Hospital, in a high-malaria area, is intended to address the gap in knowledge.

The aim of the study was to document the clinical presentation and treatment outcome of paediatric patients with SCD admitted to Rundu Hospital in 2020 and 2021.

Methods

This was a retrospective cross-sectional study using hospital records of paediatric patients aged ≤12 years with a diagnosis of SCD and admitted to Rundu Hospital from 1 January 2020 to 31 December 2021. Data were collected from 1 August to 30 September 2022. Formal consent for the study was obtained from the Ministry of Health and Social Services (MOHSS) Biomedical Research Ethics Committee (BREC) and the Research Management Committee in the Office of the Executive Director (ref. no. 22/4/2/3). Of note, the BREC is an entity under the MOHSS. The requirement for consent was waived because data were collected from the hospital records.

Data collection and analysis

Recording sheets were used to abstract the data from the hospital database for each patient. Demographic data, including age, sex and place of origin, and data on clinical presentation of SCD (crisis: vaso-occlusive, haemolytic, sequestration, aplastic), malaria illness, and serious infection were collected. Additionally, information on medication used (hydroxyurea, oral penicillin) and treatment outcome (death/discharge) was captured. An Excel spreadsheet, version 16, 2021 (Microsoft Corp., USA), was created, and data were anonymised and cleansed then transferred to SPSS, version 26.0 (IBM, USA), for analysis.

Results

Demographic characteristics of patients with SCD

Over 2-year study period (2020 - 2021) a total of 3 168 children were admitted to Rundu Hospital. Of these, 103 had SCD, which constituted a prevalence of 3.3%. The total population of children aged <15 years in the Kavango East region is 93 921, which accounts for 43% of the youth population. Of the patients with SCD, 38 (36.9%) were female and 65 (63.1%) male (Table 1). The age range was 1 - 12 years, with a median of 5 years. The majority of the children (n=56; 54.4%) were aged ≤5 years, with the remaining 35 (33.9%) and 12 (11.7%) aged 6 - 10 years and 11 - 15 years, respectively. Area of residence was categorised according to

whether the patient lived in rural or urban Kavango East. The majority of the patients (*n*=65; 63.1%) were from the urban area, with a minority (*n*=33; 32.0%) from the rural area and no clear documentation of area of residence for 5 (4.9%). All the children were Namibians, many from the Kavango tribe, but were fairly evenly distributed within the region.

Clinical presentation of patients with SCD

Most patients (*n*=64; 62.1%) presented in vaso-occlusive crisis, 28 (27.2%) presented in haemolytic crisis, and 3 (2.9%) presented in both vaso-occlusive and haemolytic crisis. Eight patients (7.8%) presented with other medical conditions, namely epilepsy, stroke, cerebral palsy, gastroenteritis and pneumonia (Fig. 1).

One patient (1.0%) had malaria, and of the total of 103 only 15 (14.6%) presented with an infection. Some of these had infection together with a crisis and some primarily had an infection. Twenty-two patients (21.4%) had multiple admissions, while 81 (78.6%) had only one admission (Table 2). None of the patients died during the study period.

Table 1. Demographic characteristics of children with sickle cell disease admitted to Rundu Intermediate State Hospital (*N*=103)

Variable	*n* (%)
Sex	
Female	38 (36.9)
Male	65 (63.1)
Age at presentation (years)	
1 - 5	56 (54.4)
6 - 10	35 (33.9)
11 - 15	12 (11.7)
Area of residence	
Urban	65 (63.1)
Rural	33 (32.0)
Missing information	5 (4.9)

The majority of the patients (*n*=90; 87.4%) received analgesics for pain, with 61 (67.9%) given an opioid. Antibiotics were administered to 99 (96.1%), the majority receiving oral penicillin (penicillin VK) and the rest ceftriaxone. Twenty-eight patients (27.2%) required a transfusion of red cell concentrate. Patients who presented in vaso-occlusive 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. We found that there were more boys 65 (63.1%) than girls 38 (36.9%). Similarly, in a study of 75 children by Mustafa *et al.*,[9] 41 (55%) were boys and 34 (45%) were girls. This male preponderance is likely to reflect differences in morbidity and mortality between the genders. In Africa, where there is a lack of newborn screening and routine childhood vaccinations, and malaria, malnutrition and poverty remain important challenges, the mortality among children with SCD who are younger than 5 years of age can be as high as 90%.[11]

Children with SCD generally start to show symptoms during the first year of life. In our study, the majority of the children with SCD (*n*=56; 54.4%) were aged ≤5 years. Similar findings were reported by Mustafa *et al.*,[9] with 52% of participants aged ≤5 years.

The majority of the patients in our study came from urban areas (63.1%) as opposed to rural areas (32.0%), with residence data missing for 4.9%. This finding may indicate that people living in an urban area have easier access to healthcare facilities compared with their rural counterparts. Income inequality, urban-rural disparities and limited access to healthcare in remote areas are persistent problems in Namibia.[22,23] These factors can delay diagnosis and treatment of SCD, especially in regions where healthcare services are scarce.

Many of the patients in our study (62.1%) presented in vaso-occlusive crisis, with 27.2% in haemolytic crisis. Painful vaso-occlusive crisis due to bony infarction is the commonest cause for hospital admission reported in other studies.[2] Infectious diseases such as malaria, pneumococcal disease, meningitis, pneumonia and septicaemia are thought to be the major cause of morbidity and mortality in SCD.[1] Of the patients in our study,

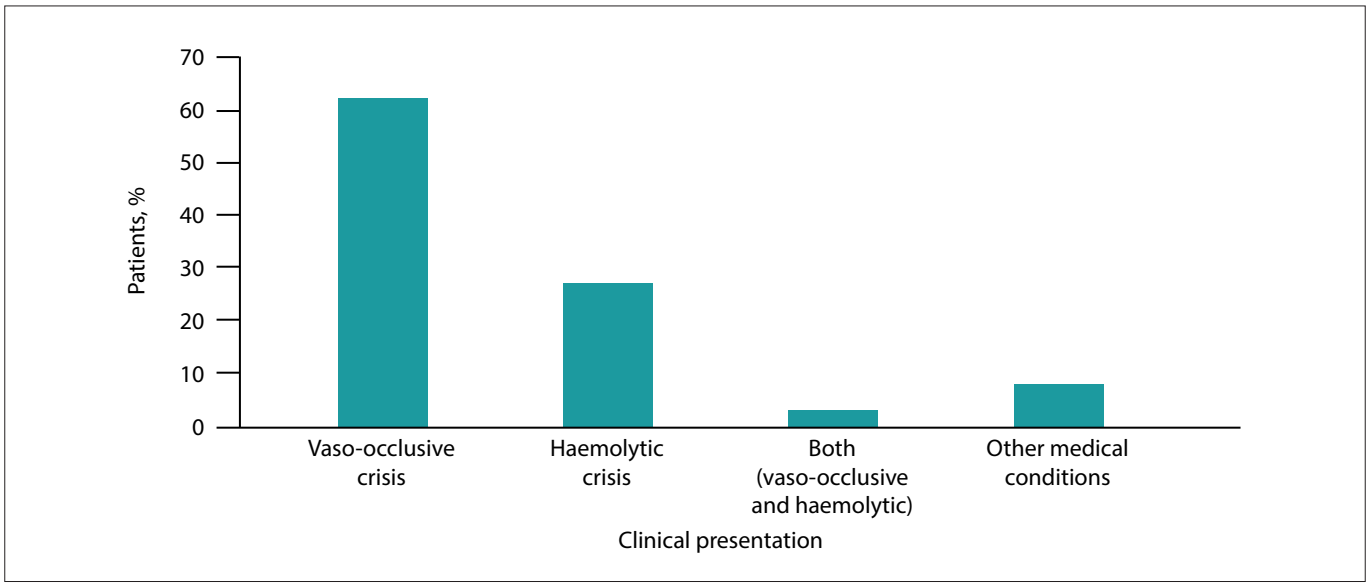

Fig. 1. Clinical presentation of patients with sickle cell disease admitted to Rundu Intermediate State Hospital (N=103).

Table 2. Malaria, other infections and multiple admissions of children with sickle cell disease (*N*=103)

Variable	*n* (%)
Malaria	
Yes	1 (1.0)
No	102 (99.0)
Infection	
Yes	15 (14.6)
No	88 (85.4)
Multiple admissions	
Yes	22 (21.4)
No	81 (78.6)

only 1 (1.0%) had malaria, and 15 (14.6%) presented with an infection. Despite the Kavango region being considered a high-risk malaria region, the diagnosis of malaria in only 1.0% of the patients may reflect success in combating malaria in Namibia.

A study conducted in a university teaching hospital in Tanzania reported an SCD mortality rate of 1.9 per 100 person-years of observation, with the highest proportion of deaths occurring in the first 5 years of life.[16] In low-resourced settings and countries where newborn screening is not yet standard care, affected children may die young even before the diagnosis is confirmed.[4]

Pain accounts for the majority of hospitalisations of patients with SCD.[4] Pain is the cardinal feature of SCD, and it is characteristically unpredictable, episodic in nature, and described as one of the most excruciating forms of pain that affect human beings.[4] It is the commonest acute complication of SCD, and significantly affects health-related quality of life.[4] Opioids and other forms of analgesia were given to our patients to alleviate pain.

SCD increases susceptibility to infections, notably bacterial sepsis and malaria in children aged <5 years.[4] Respiratory infections can trigger the sickle cell acute chest syndrome, with a high risk of death. In our study, patients with any form of infection had access to antibiotics. Additionally, essential medicines for SCD such as folic acid, oral penicillin (penicillin VK) and hydroxyurea were available to all patients. Hydroxyurea (hydroxycarbamide) remains the only agent that has been proven to reduce the number of painful crises, episodes of acute chest syndrome and hospitalisations in randomised controlled trials in adults.[2] In addition, intravenous fluids were available for the patients. Given the fact that pain is often triggered by infection, exposure to cold or dehydration, supportive care during SCD crises involves providing hydration and warmth and treating the underlying infection.[4]

Currently the only available disease-modifying medications for SCD are hydroxyurea 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 concentration for improved oxygenation in severe anaemia, and also to reduce the percentage of HbS. It may be given as a simple top-up blood transfusion or as exchange transfusion (manual or automated).[4] Twenty-eight of our patients (27.2%) received a transfusion of red cell concentrate. The main curative therapy for SCD is stem cell transplantation, while gene therapy has recently been approved by the US Food and Drug Administration.[4,24] Despite the use of disease-modifying therapy, 21.4% of our patients had multiple admissions. Definitive treatment such as gene therapy and stem cell transplantation are areas for possible future development. Moreover, early diagnosis in newborns may make it possible to intervene before the development of serious complications.

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

Conclusion

SCD remains a public health problem in Namibia, affecting 3.3% of children admitted to Rundu Hospital. Children aged ≤5 years are most commonly affected. Children with SCD presented in various crises and had access to adequate treatment. Future studies should focus on creation of registries for paediatric patients to enable proper follow-up. There is a need to improve patient record keeping to avoid gaps in future research. Additionally, a prospective study exploring future newborn screening should be considered.

Declaration. The research for this study was carried out in partial fulfilment of the requirements for CNS's MB ChB degree at the University of Namibia.

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Author contributions. CNS designed the study and drafted the article 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.

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Data availability statement. The data sets generated and analysed during the current study are available from the corresponding author (CNS) upon reasonable request.

Conflict of interest. None.

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