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Epidemiology, clinical presentation, and treatment of sickle cell disease: A retrospective study in Wangata General Referral Hospital, Mbandaka, Democratic Republic of Congo

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ABSTRACT

Introduction

Sickle cell disease (SCD) remains a major public health challenge in Mbandaka, Democratic Republic of the Congo, yet its clinical and epidemiological characteristics are insufficiently studied.

Purpose

This study aims to describe the prevalence, clinical profiles, and treatment outcomes of SCD in children treated at Wangata General Referral Hospital.

Methods

A retrospective review of medical records for children aged 2 months to 15 years hospitalised between January 2017 and December 2023 was conducted. Data were analysed using Epi Info version 7.2.5.0.

Results

Among 4,492 paediatric admissions, 64 cases of SCD were identified, yielding a prevalence of 1.4%. The case-fatality rate was 20.3%, with complications such as malaria and sepsis contributing to deaths. Hydroxyurea, a standard therapeutic agent, was not administered to any patient. The average length of hospitalisation was 3.5 days, and most deaths (53.8%) occurred within 48 hours of admission.

Conclusion

SCD in Wangata is characterised by low prevalence but high mortality, reflecting gaps in diagnosis, treatment, and hospital resources. Urgent measures are needed to improve awareness, healthcare provider training, and access to essential medications.

INTRODUCTION

Sickle cell disease (SCD) deserves attention as it causes significant challenges for households with one or more children affected by the condition (Wailoo, 2017). SCD is a public health concern in the Democratic Republic of Congo (DRC) and represents a considerable burden (Tshilolo et al., 2009). It is one of the major public health issues in the DRC (Shongo ya Pongombo, 2022). The population of Mbandaka is also affected by SCD. A recent study in Mbandaka revealed a prevalence of 22.99% for carriers of the sickle cell trait (Eleke Ebola et al., 2024).

However, health facilities in Mbandaka focus exclusively on the care of patients with SCD but do not examine its epidemiology, impact, or associated challenges. This study aims to describe the prevalence, clinical profiles, and treatment outcomes of SCD in children treated at Wangata General Referral Hospital (Wangata GRH).

METHODS

A retrospective, descriptive cross-sectional study was conducted using patient records from Wangata GRH, Mbandaka, DR Congo, between January 1, 2017, and December 31, 2023. The study included children aged 2 months to 15 years diagnosed with SCD. Inclusion criteria required complete medical records with key clinical and demographic variables, while patients with incomplete data were excluded.

Data Collection and Variables

The study examined variables such as patient age, sex, reasons for hospitalisation, laboratory investigations, treatment regimens, length of stay, and outcomes (survival or death). The presence of comorbid conditions and financial support for healthcare costs were also recorded.

Statistical Analysis

Data were entered into Microsoft Excel and analysed using Epi Info version 7.2.5.0. Descriptive statistics, such as frequencies and percentages, were calculated for categorical variables. Continuous variables (e.g., age, length of hospitalisation, and periods for hospital service requests) were summarised as means.

Ethical Considerations

The research protocol for this study was submitted to and approved by the Ethics Committee of the Higher Institute of Medical Techniques of Kinshasa, Democratic Republic of the Congo. This was a retrospective study. Access to children's files was authorised by the hospital's medical director. Anonymity was strictly observed. In the database, the children's names were coded and replaced by numbers.

RESULTS

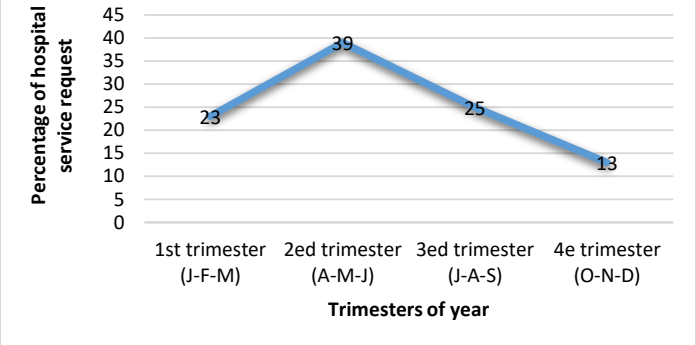
The results are divided into three sections. The first section addresses the epidemiology of SCD at this hospital. The second focuses on the clinical aspects and diagnosis. The third examines financial support for the care of children with SCD.

Epidemiology of Sickle Cell Disease

Among 4,492 paediatric admissions, 64 cases of SCD were identified, yielding a prevalence of 1.4%. Of these, 34 were girls (53.1%) and 30 were boys (46.9%). Twenty-two patients (34.3%) were under 5 years of age. The case-fatality rate was 20.3%. The average length of hospitalisation was 3.5 days, with most deaths (53.8%) occurring within 48 hours of admission.

Requests for hospital services occurred throughout the year. The rate of requests for hospital services by children with SCD was 23% in the first quarter, 39% in the second quarter, 25% in the third quarter, and 13% in the last quarter of the year. The average quarterly rate of demand for hospital services was 16%.

Figure 1:
Annual evolution of hospital service requests by children with SCD



Sickle Cell Disease Clinic

The results in this section relate to the following areas:

- Diagnosis of sickle cell disease
- Length of hospital stay for patients
- Use of hydroxyurea in patient treatment

- Outcome of patient hospitalisation

Diagnosis of Sickle Cell Disease

Clinical and paraclinical diagnosis of SCD was inadequate. Confirmatory paraclinical diagnosis of SCD is essential for proper management, but in this hospital, the Emmel test was the only available test for sickle cell screening. No child suspected of having SCD received a confirmatory diagnosis using a more advanced test.

Although 32 patients (50%) were referred to the laboratory for paraclinical testing for SCD, only 14 (21.9%) returned with test results. This highlights a significant gap in diagnostic follow-up.

Length of Stay in Hospital

The average length of hospital stay was 3.5 days. The distribution of hospital stays is as follows:

- 1 to 2 days: 40.6% of cases
- 3 to 5 days: 34.4% of cases
- More than 5 days: 25% of cases

This indicates that the majority of patients (75%) were hospitalised for less than 5 days.

Use of Hydroxyurea in Patient Treatment

No child with SCD received hydroxyurea as part of their treatment (0%). None of the children had hydroxyurea included in their medical prescriptions. This finding points to a treatment gap, as hydroxyurea is a standard treatment for SCD (Tshilolo et al., 2009).

Outcome of Hospitalisation and Pathologies Responsible for Death

Out of the 64 children with SCD, 51 (79.7%) were discharged alive, while 13 (20.3%) died. Twenty-three pathologies were identified as contributing to the deaths. Among these, SCD complications were responsible for 44% of deaths, while malaria was responsible for 17% of deaths. Other contributing conditions included meningitis (9%), sepsis with a pulmonary or digestive portal (9%), leukaemia (9%), unspecified mesenteric crisis (4%), hypoglycaemia (4%), and kidney complications (4%). Comorbidity was notably high, with a prevalence of 56% among deceased patients.

Table 1:
Distribution of children who died in hospital according to the pathologies involved

Pathology involved with the death of children with SCD	Number of deaths	%
Sickle cell disease (complications)	10	44
Malaria	4	17
Meningitis	2	9
Sepsis (pulmonary or digestive portal)	2	9
Leukaemia	2	9
Unspecified mesenteric crisis	1	4
Hypoglycaemia	1	4
Kidney complication	1	4
Total	**23**	**100**

This distribution underscores the burden of co-existing pathologies in SCD patients.

Financing Medical Care for Children with Sickle Cell Disease

Of the 64 children with SCD, 18 (28.1%) received additional financial support from their parents' employers or external organisations. This indicates that most families had to bear the financial burden of care, which may have affected healthcare-seeking behaviours (Eleke et al., 2024).

DISCUSSION

The prevalence of sickle cell disease (SCD) in this hospital was low (1.4%) compared with those reported by other hospital-based studies. At the Sikasso hospital in Mali and a hospital in Uganda, prevalences of 1.67% (Ouédraogo et al., 2014) and 1.67% (Cissouma et al., 2021) were noted, respectively.

This study was characterised by a slight female predominance (53.1%). This result aligns with findings from other studies, which reported a representation of girls at 60.9% (Koko et al., 1998) and 51% (Chemegni et al., 2018), respectively.

Compared with the lethality of SCD reported in health establishments—2.5% at the Centre National Hospitalier Universitaire-Hubert Koutoukou Maga in Cotonou, Benin (Dodo et al., 1998) and 3.1% at the Centre Hospitalier Régional de Saint-Louis in Senegal (Seck et al., 2021)—the lethality observed in this study was considerably higher. This suggests that the care given to sickle cell patients at the Wangata General Referral Hospital (HGR) is inadequate and requires further investigation. This can also be attributed to a lack of awareness of SCD among children's parents and healthcare providers (Dahmani et al., 2017).

Children over the age of 5 admitted to the paediatric ward of the Wangata HGR were more represented (65.6%) than those under the age of 5. This is understandable, given that the hospital does not have a high-performance technical platform to detect all cases of sickle cell disease. Most of the children in this study were survivors of sickle cell crises from the first five years of the disease's manifestation. The literature indicates that many children with SCD do not survive beyond their fifth birthday. In countries with a high prevalence of SCD, under-five mortality associated with SCD ranges from 9% to 16% (Odièvre, 2022).

Deaths among sickle cell children in this study were mainly due to complications of SCD (44%). General comorbidity and the presence of malaria also played a significant role. Deaths associated with SCD can be explained by the pernicious nature and unpredictable course of the disease (Wailoo, 2017). Sickle cell disease often worsens as the immune system weakens (Tshilolo & Aissa, 2009).

Mbandaka is located in a permanent malaria transmission area, which is why malaria has played such a major role. Malaria exacerbates the condition of sickle cell patients, who naturally have a lower haemoglobin level than their healthy peers, causing further anaemia.

Hospital service requests for sickle cell children were predominantly high in the second half of the year (39%) but low in the final half (13%). This pattern was characterised by seasonal variations. Several factors could explain this variability, including comorbidity, climatic effects (particularly the cold during rainy periods), and dehydration observed during the dry season. Some authors have associated this variability with atmospheric pollutants (Tewari et al., 2015).

Early diagnosis and treatment can prevent complications (Tshilolo & Aissi, 2009). However, the rates of request (50%) and completion of paraclinical examinations for SCD (21.9%) were low. Most children (53.8%) died within 48 hours of being admitted to the hospital. This suggests a delay in seeking care. Compared with Mbujimayi and Lubumbashi, where 88% of patients received prescriptions for hydroxyurea (Mbiya et al., 2021), those admitted to the Wangata HGR were not treated with hydroxyurea, and this drug was not included in their medical prescriptions. Hydroxyurea is now recognised as a drug that significantly improves the clinical and biological parameters of SCD and reduces the risk of malaria and infectious complications (Aliyu et al., 2007; Cally et al., 2015). A correct diagnosis can reduce the hospital case-fatality rate and complications of SCD (Bitwe et al., 2009).

The management of sickle cell disease at the Wangata HGR suffers from significant shortcomings, which jeopardise the prognosis of children with the disease. The hospital's sickle cell management was limited as it had neither haemoglobin electrophoresis for the confirmatory diagnosis of SCD nor hydroxyurea. Moreover, healthcare providers were not sufficiently trained in the management of SCD. This was evidenced by the lack of systematic paraclinical tests for SCD and the absence of medical prescriptions for hydroxyurea.

Compared to the average length of hospitalisation of 4.5 days observed at a hospital centre in Benin (Dodo et al., 2018), the average hospitalisation period at Wangata HGR was relatively short, at 3.5 days. Financial support for sickle cell patients was rare, with only 28.1% of patients receiving additional financial support for care. Financial support is essential for SCD patients, as they often require medical care due to their predisposition to illness.

CONCLUSION

Sickle cell patients seek care at Wangata General Referral Hospital year-round, yet the hospital faces significant challenges in managing SCD cases. Despite a low prevalence of 1.4%, the hospital recorded a high lethality rate. Deaths were primarily linked to SCD complications and co-morbidities, compounded by limited diagnostic and therapeutic resources. Efforts should focus on strengthening hospital infrastructure, increasing access to hydroxyurea, improving diagnostic capacity, and raising community awareness to reduce mortality.

Recommendations

An urgent effort is needed at Wangata HGR to organise training for healthcare providers and to procure diagnostic equipment for the confirmatory diagnosis of SCD as well as hydroxyurea. Pregnant women also need to be sensitised about sickle cell anaemia, just as they are about other diseases.

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Conflicts of Interest: None declasred.

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