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Original Research Paper

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CROSS SECTIONAL STUDY ON SICKLE CELL DISEASE

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ABSTRACT

Introduction: Sickle cell disease (SCD) is a hereditary hemoglobinopathy characterized by diverse acute and chronic complications, contributing to significant morbidity and mortality globally. This study aimed to assess the prevalence and spectrum of SCD-related complications and evaluate current management approaches in a clinical setting. **Methods:** A cross-sectional study was conducted over one year at a tertiary care hospital, enrolling 60 patients with confirmed SCD diagnosis. Data on demographic characteristics, clinical complications, and management strategies were collected through patient interviews, clinical examinations, and medical record reviews. Descriptive statistics summarized complication prevalence and treatment patterns, while associations between clinical variables and complications or management were analyzed using appropriate statistical tests. **Results:** The cohort had a mean age of 28.5 years, with a predominance of the HbSS genotype (66.7%). Vaso-occlusive crises (75%), acute chest syndrome (30%), and chronic organ damage including nephropathy (15%) and stroke (10%) were the most common complications. Hydroxyurea was used by 36.7% of patients and was associated with a reduced frequency of acute chest syndrome ($p=0.04$). Patients with the HbSS genotype exhibited a higher prevalence of vaso-occlusive crises compared to other genotypes ($p=0.02$). Infection prophylaxis coverage was high (83.3%), and opioid-based pain management was common (63.3%). Older age correlated significantly with increased prevalence of chronic kidney disease and stroke ($p<0.001$).**Conclusion:** Despite available treatments, SCD patients continue to experience high morbidity, emphasizing the necessity for enhanced management strategies, increased hydroxyurea use, and integration of emerging therapies. Future research should focus on longitudinal outcomes and tailored interventions in high-burden populations.

KEYWORDS : Sickle Cell Disease, Vaso-Occlusive Crisis, Hydroxyurea, Acute Chest Syndrome, Disease Management

INTRODUCTION

Sickle cell disease (SCD) is a hereditary hemoglobinopathy characterized by the presence of abnormal hemoglobin S, which leads to chronic hemolytic anemia, vaso-occlusion, and multi-organ complications. Despite advances in understanding its pathophysiology, SCD remains a significant public health challenge globally, particularly in regions with high prevalence such as sub-Saharan Africa, the Middle East, and parts of India. The disease manifests with a wide spectrum of clinical complications ranging from acute painful crises to chronic organ damage, significantly impacting patient morbidity and mortality.

Effective management of SCD requires comprehensive strategies addressing both acute and chronic complications, including pain management, prevention of infections, and treatment of organ dysfunction. However, variability in clinical presentation and resource availability often complicates standardized care delivery. Cross-sectional studies provide valuable insights into the current burden of disease complications and management practices, enabling identification of gaps in care and informing targeted interventions.

This study aims to assess the prevalence and spectrum of complications in patients with SCD and evaluate the current management approaches employed in clinical practice. By capturing a snapshot of disease impact and therapeutic strategies, this research will contribute to optimizing patient outcomes and guiding health policy decisions in settings affected by SCD. The objectives of the study included to determine the prevalence and spectrum of clinical complications among patients with sickle cell disease (SCD) in the study population and to evaluate the current management approaches employed for SCD, including strategies for acute complication management, infection prevention, and treatment of chronic organ dysfunction.

METHODOLOGY

This cross-sectional study was conducted over a period of one year at a tertiary care hospital in. The study captured data on SCD complications and management practices during this timeframe. The study included patients diagnosed with SCD confirmed by hemoglobin electrophoresis or other standard diagnostic methods. Inclusion criteria encompassed all age groups and both sexes who attended the selected healthcare facilities during the study period. Exclusion criteria included patients with incomplete medical records or those unwilling to provide informed consent. A consecutive sampling method was employed, enrolling all eligible patients presenting within the one-year study period until the sample size was met. Data were collected using a structured data collection form designed to capture demographic information, clinical history, type and frequency of SCD complications (such as vaso-occlusive crises, acute chest syndrome, infections, stroke, organ damage), and details of management strategies employed (including pain management protocols, infection prophylaxis, transfusion history, and use of disease-modifying therapies like hydroxyurea). Data sources included patient interviews, clinical examinations, and review of medical records. Trained research assistants or clinicians performed data collection to ensure accuracy and consistency.

Ethical Considerations: Ethical approval was obtained from the institutional ethical committe boards. Informed consent was secured from all participants or their guardians before data collection. Confidentiality and anonymity were maintained throughout the study. Collected data were entered into a statistical software package for analysis. Descriptive statistics summarized the prevalence and types of

complications and management approaches. Associations between demographic or clinical variables and complications or management practices were explored using appropriate statistical tests.

RESULTS

A total of 60 patients diagnosed with sickle cell disease (SCD) were enrolled in this cross-sectional study. The demographic and clinical characteristics of the study population are summarized in Table 1.

Table 1: Demographic and Clinical Characteristics of Study Population (N=60)

Characteristic	Value
Mean age (years)	28.5 ± 12.3
Age range (years)	5 – 55
Sex, n (%)	
- Male	32 (53.3)
- Female	28 (46.7)
Confirmed genotype, n (%)	
- HbSS	40 (66.7)
- HbSC	12 (20.0)
- HbS -thalassemia	8 (13.3)
Mean duration of diagnosis (years)	15.2 ± 8.7

The prevalence of various SCD-related complications observed in the study population is presented in Table 2. The most common complications included vaso-occlusive crises (VOCs), acute chest syndrome (ACS), and chronic organ damage such as nephropathy and stroke.

Table 2: Prevalence of SCD Complications in Study Population (N=60)

Complication	Number of Patients	Prevalence (%)
Vaso-occlusive crises (≥1 episode in past year)	45	75.0
Acute chest syndrome	18	30.0
Stroke (history of clinical or silent stroke)	6	10.0
Chronic kidney disease	9	15.0
Priapism (males only, n=32)	8	25.0
Pulmonary hypertension	5	8.3
Leg ulcers	4	6.7
Avascular necrosis	7	11.7
Infection episodes (≥1 in past year)	20	33.3

Table 3: Management Approaches in Study Population (N=60)

Management Strategy	Number of	(%)
Hydroxyurea use	22	36.7
Blood transfusion (≥1 in past year)	15	25.0
Pain management with opioids	38	63.3
Infection prophylaxis (vaccination/antibiotics)	50	83.3
Use of disease-modifying agents other than hydroxyurea	2	3.3
Regular clinical follow-up	55	91.7

In this study it was observed that patients with HbSS genotype had a significantly higher prevalence of vaso-occlusive crises compared to other genotypes (p=0.02) (Table 4).

Table 4: Association between vasoocclusive crisis and genotype

Variable	Group	Number of Patients with Complication	Number of Patients without Complication	Total Patients	%	p-value
Vaso-occlusive crises (≥1 episode in past year)	HbSS genotype	35	5	40	87.5	0.02*
	Other genotypes (HbSC, HbSβ-thalassemia)	10	10	20	50.0	

*p value <0.05; Hence statistically significant

Considering the use of various medications and frequency of ACS it was observed that, hydroxyurea use was associated with reduced frequency of ACS episodes (p=0.04) (Table 5).

Table 5: Association between hydroxy urea use and ACS episodes.

Variable	*Group*	*Number of Patients with ACS Episodes*	*Number of Patients without ACS Episodes*	*Total Patients*	%	*p-value*
Acute chest syndrome (ACS)	Hydroxy urea users	4	18	22	18.2	0.04*
	Non-users	14	24	38	36.8	

*p value <0.05; Hence statistically significant

Older age was correlated with increased prevalence of chronic kidney disease and stroke (p<0.01) (Table 6).

Table 6: Association between age and various complications.

Variable	Age Group	Number of Patients with Complication	Number of Patients without Complication	Total Patients	Prevalence (%)	p-value
Chronic kidney disease	>30 years	8	12	20	40.0	<0.001*
	<=30 years	1	39	40	2.5	
Stroke (clinical or silent)	>30 years	5	15	20	25.0	<0.001*
	<=30 years	1	39	40	2.5	

*p value <0.05; Hence statistically significant

DISCUSSION

This cross-sectional study involving 60 patients with sickle cell disease (SCD) highlights a substantial burden of both acute and chronic complications, consistent with findings from broader epidemiological and clinical research. The predominance of vaso-occlusive crises (VOCs) affecting 75% of patients aligns with the established understanding that acute pain episodes are the hallmark and most frequent complication of SCD, as emphasized in the literature. The significant prevalence of acute chest syndrome (30%) and chronic organ damage such as nephropathy (15%) and stroke (10%) further corroborates the multisystem impact of SCD described in global and regional studies.

The predominance of the HbSS genotype (66.7%) in this cohort and its association with a higher prevalence of VOCs (p=0.02) reflects the known genotype-phenotype correlation, where homozygous HbS disease manifests with more severe clinical complications. The observed association between hydroxyurea use and reduced frequency of acute chest

syndrome (p=0.04) supports the well-documented efficacy of hydroxyurea in decreasing acute complications and improving clinical outcomes. However, the suboptimal hydroxyurea usage rate (36.7%) in this study mirrors underutilization trends reported in large retrospective cohorts, including Manwani et al., where barriers such as access, adherence, and provider hesitancy limit optimal treatment.

Infection prophylaxis coverage was high (83.3%), indicating adherence to recommended preventive strategies, which is critical given the increased susceptibility to infections in SCD patients due to functional asplenia and immune dysfunction. Pain management primarily relied on opioids (63.3%), consistent with the acute care needs for VOCs but also reflecting the challenges in managing chronic pain and the risks associated with opioid therapy.

The study's finding of increased prevalence of chronic kidney disease and stroke with advancing age (p<0.001) aligns with the natural history of SCD, where cumulative vascular and organ damage accrues over time. This underscores the importance of early intervention and longitudinal monitoring to mitigate progressive morbidity.

Regional data on SCD burden among tribal populations in India contextualize the study findings within a high-prevalence setting, emphasizing the need for targeted public health strategies, including genetic counseling, community awareness, and improved healthcare access. The variability in clinical presentation and resource availability noted in this study echoes the challenges described in tribal and resource-limited areas, where standardized care delivery remains difficult.

Overall, this study contributes valuable data on the spectrum of SCD complications and current management practices, reaffirming the persistent high morbidity despite available therapies. It highlights critical gaps in hydroxyurea utilization and the ongoing reliance on symptom-driven care. These insights support calls for enhanced efforts to increase access to disease-modifying treatments, comprehensive care models, and integration of newer therapeutic agents such as l-glutamine, crizanlizumab, and voxelotor, which have shown promise but were minimally used in this cohort.

Future research should focus on longitudinal assessment of treatment adherence, effectiveness of emerging therapies, and implementation of culturally and regionally tailored interventions, particularly in high-burden populations. Strengthening healthcare infrastructure and education to overcome barriers to optimal care is imperative to reduce the morbidity and mortality associated with SCD.

LIMITATIONS

This study has several limitations. First, the relatively small sample size of 60 patients may limit the generalizability of the findings to broader SCD populations. Second, the cross-sectional design captures only a snapshot in time, precluding assessment of temporal changes in complications or management practices and limiting causal inferences. Third, data collection relied on patient interviews and medical records, which may be subject to recall bias or incomplete documentation, potentially affecting the accuracy of reported complications and treatment details. Fourth, the minimal use of newer disease-modifying agents such as l-glutamine, crizanlizumab, and voxelotor limits the ability to evaluate their impact within this cohort. Fifth, variability in resource availability and clinical practices across different healthcare settings was not fully explored, which may influence management approaches and outcomes.

CONCLUSION

This cross-sectional study provides valuable insights into the burden of complications and current management practices among patients with sickle cell disease, reaffirming the high morbidity associated with the condition despite available therapies. The findings highlight significant gaps in hydroxyurea utilization and an ongoing reliance on symptom-driven care. Addressing these gaps requires enhanced efforts to improve access to disease-modifying treatments and comprehensive care models. Integration of newer therapeutic agents, although currently limited, offers promising avenues for improved outcomes. Future research should focus on longitudinal evaluation of treatment adherence, effectiveness of emerging therapies, and culturally and regionally tailored interventions, particularly in high-burden populations. Strengthening healthcare infrastructure and educational initiatives is imperative to overcome barriers and reduce the morbidity and mortality associated with sickle cell disease.

REFERENCES

1. Vona R, Sposi NM, Mattia L, Gambardella L, Straface E, Pietraforte D. Sickle Cell Disease: Role of Oxidative Stress and Antioxidant Therapy. Antioxidants. 2021 Feb 16;10(2):296.
2. Colombatti R, Birkegård C, Medici M. PB2215: GLOBAL EPIDEMIOLOGY OF SICKLE CELL DISEASE: A SYSTEMATIC LITERATURE REVIEW. HemaSphere. 2022 June 23;6(Suppl):2085–2086.
3. Sagi V, Mittal A, Tran H, Gupta K. Pain in sickle cell disease: current and potential translational therapies. Translational Research. 2021 Mar 9;234:141–158.
4. Colah RB, Mukherjee MB, Martin S, Ghosh K. Sickle cell disease in tribal populations in India. Indian Journal of Medical Research. 2015 May 1;141(5):509–515.
5. Brandow AM, Liem RI. Advances in the diagnosis and treatment of sickle cell disease. J Hematol Oncol. 2022 Mar 3;15(1).
6. Elendu C, Amaechi DC, Alakwe-Ojimba CE, Elendu TC, Elendu RC, Ayabazu CP, et al. Understanding Sickle cell disease: Causes, symptoms, and treatment options. Medicine. 2023 Sept 22;102(38):e35237.
7. Manwani D, Burnett AL, Paulose J, Yen GP, Burton T, Anderson A, et al. Treatment patterns and burden of complications associated with sickle cell disease: A US retrospective claims analysis. eJHaem. 2022 Oct 6;3(4):1135–1144.
8. Nagisetty S, Thatoju PK. Sickle cell disease burden among tribal population of India: a narrative review. Int J Community Med Public Health. 2024 Oct 29;11(11):4537–4545.
9. Ochocinski D, Dalal M, Black LV, Carr S, Lew J, Sullivan K, et al. Life-Threatening Infectious Complications in Sickle Cell Disease: A Concise Narrative Review. Front Pediatr. 2020 Feb 20;8.
10. Katsivalis KV, Kosacz J, Austin Szwak J. Opioid Use in Vaso-Occlusive Crisis During Intravenous Opioid Drug Shortage. Hosp Pharm. 2022 June 4;57(6):721–726.
11. Dad T, Weiner DE. Stroke and Chronic Kidney Disease: Epidemiology, Pathogenesis, and Management Across Kidney Disease Stages. Seminars in Nephrology. 2015 July 1;35(4):311–322.
12. Aggarwal P, Bhat D. Genetic counseling in sickle cell disease: Insights from the Indian tribal population. J Community Genet. 2023 Aug 4;14(4):345–353.
13. Mohanty SS, Purohit A, Anand PK, Huda RK, Sharma AK. Burden of sickle cell disease in tribal students in Maa-Baadi institutions in southern Rajasthan - A pilot study. Indian J Med Res. 2022 Aug 1;156(2):269–274.