

Original Article

Epidemiology of vaso-occlusive crisis among sickle cell disease patients in India: A community-based multi-centric study

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Background and objectives: Morbidity burden of sickle cell disease (SCD) in India has been underestimated till now due to a lack of nationwide data on the prevalence of clinical outcomes, including the most prevalent clinical manifestations like vaso-occlusive crises (VOCs). This multi-centric study reports the epidemiological characteristics of VOCs among SCD patients in India.

Methods: This is a prospective, community-based, multi-centre cohort study with fortnightly home visits conducted over 12 months in five SCD-endemic districts of India. A cohort of 252 patients was followed up, and data regarding any illness related to SCD and its management in the previous fortnight were collected using a structured questionnaire.

Results: Out of the total 252 SCD patients followed up, 223 (88.5%) patients reported 2118 crises in a year [8404 episodes per 1000 patients-year; 95% confidence interval (CI): 8040-8760]. Pain anywhere in the body was the most prevalent symptom [97.3% (n=217) of the participants, with 7 (3-13) median (IQR) episodes per patient in a year]. Among total crisis episodes, 48.7% (n=1031) of the episodes were treated at a health facility.

Interpretation and conclusions: A higher number of crisis episodes reported among the majority of the patients demystifies the notion that SCD is milder and less severe in India. The lower utilization of healthcare resources among SCD patients raises a serious concern.

Keywords Acute illness; Epidemiology; Sickle cell disease; Tribal health; Vaso-occlusive crisis

Sickle cell disease (SCD), a monogenic disorder caused by a single-nucleotide mutation, is the second most common hemoglobinopathy after thalassemia in India, prevalent especially among the tribal, an underserved population.^{1,2} Clinical manifestations of SCD vary from asymptomatic to mild to severe forms associated with high morbidity and mortality.³ It is primarily presented with symptoms of anaemia, vaso-occlusive crisis (VOC), splenic sequestration, avascular bone necrosis, and osteomyelitis.⁴⁻⁶

The causative mutation of SCD in India is primarily associated with the Arab Indian haplotype, characterised

by a relatively high percentage of foetal haemoglobin (HbF), which is considered mild compared to African haplotypes.⁷⁻⁹ The country's diverse landscape also contributes to the heterogeneous presentation of SCD in India.^{10,11} However, the severity and variability of the disease have been less well understood in India.^{10,12} Recent epidemiological studies in India revealed a high prevalence of severe SCD phenotype despite high HbF levels.^{11,13-15}

There is a dearth of information about many aspects of SCD in Indian patients, including the natural history and prevalent clinical manifestations

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like VOC.¹⁶ The natural history of the disease in India remains unestablished. Most of our knowledge and understanding of the natural history of SCD comes from developed countries, where effective interventions are available.¹⁷ A few studies conducted in India, based at a single institution or region, are inconclusive and underestimate the burden.¹⁸⁻²⁰ Nationwide studies on the epidemiology of VOCs are also very few.²¹ And this situation limited the development of locally appropriate models of clinical care.²²

The VOCs and their management significantly impact the clinical burden, QoL, and healthcare utilisation among SCD patients.^{23,24} Therefore, there is a dire need to study the epidemiology of SCD-related clinical manifestations across India. A prospective, community-based, multi-centre study with frequent (fortnightly) follow-up is better suited to capture real-world episode occurrence, care pathways, and site-level heterogeneity across underserved districts. This design addresses gaps in the Indian literature and complements hospital-based studies by documenting VOCs irrespective of place of care. The present study was conducted to document the epidemiology of VOCs through fortnightly visits over one year among SCD patients across five SCD-endemic districts in India.

Methods

This prospective study was undertaken together by the department of Biotechnology, Central Tribal University of Andhra Pradesh, Vizianagaram, Andhra Pradesh; department of Community Medicine, Parul Institute of Medical Sciences and Research, Vadodara, Gujarat; division of Molecular Epidemiology, ICMR-Regional Medical Research Centre, Bhubaneswar, Odisha; department of Anatomy, JSS Medical College, Mysore, Karnataka; and department of Biotechnology, Bodoland University, Kokrajhar, Assam. The study was approved by the Institutional Ethics Committees of all participating institutions corresponding to the respective study districts. Informed consent was obtained from all adult participants. For participants below 18 years of age, informed consent was obtained from a parent or legal guardian, along with age-appropriate assent.

Study design and setting: We conducted a prospective, community-based, multi-centre cohort study across five SCD-endemic districts in India - Alluri Seetharama Raju (Andhra Pradesh), Chhotaudepur (Gujarat), Kandhamal (Odisha), Mysuru (Karnataka), and Udalguri (Assam), during the year 2023-24. These districts represent regions with documented high

HbS gene frequencies, based on publicly available estimates from the website of the National Sickle Cell Anaemia Elimination Mission (NSCAEM) (<https://sickle.nhm.gov.in/>). Further details of these districts are available elsewhere.²⁵ In these districts, healthcare is delivered through a network of public primary, secondary, and tertiary healthcare facilities. Primary healthcare facilities provide routine care, basic management of acute pain episodes, counselling, and follow-up, while patients requiring hospitalisation or blood transfusion are referred to tertiary-level hospitals. Programmatic provisions include community awareness activities and the prescription and dispensing of hydroxyurea at the primary care level, although access to timely acute care and continuity of treatment varies across districts.²⁶

SCD patients were recruited from the Indian Sickle Cell Disease Registry (ISCDR), established by the Indian Council of Medical Research in the above-mentioned districts.²⁷ The registry prospectively captured demographic, clinical, treatment, and healthcare utilisation data for SCD patients through an android-based application. It has been integrated with a three-tier screening and referral system.

Study participants and sampling: SCD patients were recruited from the two primary health centers' areas in each of the districts. We identified these patients from the registry described above for another intervention study conducted by us.²⁸ For the current study, these patients with confirmed SCD (N=261) were purposively recruited. For patients under 18 yr old, their parents or immediate caregivers were recruited. This sample size was finalised after excluding unwilling participants, those who had migrated, or those who had passed away due to the disease. Of 261 patients, 252 were followed up through the end of the study. Inclusion criteria were a laboratory-confirmed diagnosis of SCD, residence within the study area, and willingness to participate in periodic follow up. No specific intervention was introduced immediately before or during the follow up period. One year after the initiation of follow up, programmatic strengthening efforts to improve routine SCD care had been implemented. Regarding precision, this was a pragmatic cohort determined by enrolment capacity across sites during the study window; all available, consenting registry patients were included. For transparency, we report achieved precision: with n=252, the proportion of participants with ≥ 1 VOC in 12 months has an approximate 95% CI half-width of about ± 4.0 percentage points ($SE \approx 0.020$), supporting stable site- and subgroup-wise estimates. The achieved

sample size provided acceptable precision for key estimates.

Because participants were pre-identified as SCD cases purposively sampled from the cohort, the index date (time zero) was the date of cohort enrolment (June 1, 2023), and the first patient visit was on June 15, 2023. Follow up was scheduled every ~15 days for 12 months from the index date; participants were censored at 12 months, death, or last contact. Time was measured as days from the index date. This pragmatic cohort reflects programmatic feasibility, with interpretability supported by the achieved precision in reporting.

Data collection: Data were collected by a quantitative questionnaire designed and validated during the study. The study questionnaire was initially developed in English through a literature review and brainstorming among subject experts. Later, they were translated into different regional languages. These versions of the questionnaires were pre-tested, and the final versions were developed.

All study participants were visited fortnightly by the research team for one year. Thus, 24 fortnightly visits were conducted for each patient. This questionnaire was administered to patients who reported any SCD-related illness episode in the past fortnight. It included variables such as symptoms observed during the illness, the frequency and duration of these episodes, healthcare, and hospitalisation. Care was taken to differentiate VOC and chronic pain commonly seen in SCD. Necessary guidelines were developed.²⁹ VOCs and their management-related data were self-reported and verified when medical records were available. VOCs and their management details were recorded, regardless of the care setting. Socio-demographic characteristics of the study participants were collected during enrolment.

At each home visit, trained staff administered a structured questionnaire with a 15-day recall window, chosen to balance recall accuracy and operational feasibility. Tools were translated and pre-tested, field teams were standardised, and quality assurance included spot checks and supervisory review, ensuring consistency across sites.

Statistical analysis: Data were managed and analysed using SPSS, version 27 (IBM Corp., Armonk, NY, USA). Socio-demographic characteristics, SCD-related symptoms, their frequency, and duration are presented using descriptive statistics. Categorical variables are presented as frequencies and percentages. Frequencies and percentages are presented with corresponding 95%

Table I. Socio-demographic characteristics of study participants (N=252)

Variables	Number (%)
Age	
Children (≤ 10 yr)	58 (23.0)
Adolescents (11-18 yr)	70 (27.8)
Young adults (19-24 yr)	31 (12.3)
Adults (25-44 yr)	68 (27.0)
Middle-aged (45-59 yr)	20 (7.9)
Old aged (≥ 60 yr)	5 (2.0)
Gender	
Male	106 (42.1)
Female	146 (57.9)
Primary occupation	
Agriculture/Agriculture labour/ Gathering and hunting	48 (19.0)
Paid job	12 (4.8)
Homemaker	32 (12.7)
Student/Presently not working	160 (63.5)
Education	
No formal schooling	43 (17.1)
1-5 yr of schooling	71 (28.2)
6-10 yr of schooling	97 (38.5)
>10 yr of schooling	41 (16.3)
Annual family income (in INR)	
Up to 20,000	39 (15.5)
20,001 to 40,000	42 (16.7)
40,001 to 60,000	50 (19.8)
>60,000	121 (48.0)
Regular source of medical care	
Public healthcare system	201 (79.8)
Private healthcare system	26 (10.3)
Others	25 (9.9)
Hydroxyurea consumption	
Consumed regularly	6 (2.4)
Consumed intermittently	232 (92.1)
Never consumed	14 (5.5)

confidence intervals (CI) to indicate the precision of the estimates. Continuous variables are presented as means and standard deviations, or medians and interquartile ranges, based on their distribution.

Results

Socio-demographic characteristics of patients: Socio-demographic characteristics of study participants (n=252) are presented in **Table I**.

Table II. Annual incidence of vaso-occlusive crisis episodes among different age groups in males and females

Gender	Age group (yr)	Number of episodes					
		0 n (%)	1-5 n (%)	6-10 n (%)	11-15 n (%)	16-20 n (%)	>20 n (%)
Male	0-10 (n=24)	4 (16.7)	13 (54.2)	3 (12.5)	2 (8.3)	1 (4.2)	1 (4.2)
	11-20 (n = 40)	4 (10.0)	18 (45.0)	12 (30.0)	3 (7.5)	1 (2.5)	2 (5.0)
	21-30 (n=13)	1 (7.70)	3 (23.1)	2 (15.4)	3 (23.1)	3 (23.1)	1 (7.7)
	31-40 (n=16)	2 (12.5)	3 (18.7)	7 (43.7)	1 (6.2)	1 (6.2)	2 (12.5)
	41-50 (n=8)	2 (25.0)	2 (25.0)	0 (0.0)	3 (37.5)	1 (12.5)	0 (0.0)
	51 & above (n=5)	2 (40.0)	0 (0.0)	3 (60.0)	0 (0.0)	0 (0.0)	0 (0.0)
Female	0-10 (n=34)	5 (14.7)	14 (41.2)	7 (20.6)	6 (17.6)	1 (2.9)	1 (2.9)
	11-20 (n=45)	4 (8.9)	13 (28.9)	12 (26.7)	10 (22.2)	3 (6.7)	3 (6.7)
	21-30 (n=28)	2 (7.1)	9 (32.1)	10 (35.7)	2 (7.1)	1 (3.6)	4 (14.3)
	31-40 (n=20)	2 (10.0)	3 (15.0)	6 (30.0)	5 (25.0)	1 (5.0)	3 (15.0)
	41-50 (n=13)	1 (7.7)	4 (30.8)	3 (23.1)	3 (23.1)	1 (7.7)	1 (7.7)
	51 & above (n=6)	0 (0.0)	2 (33.3)	0 (0.0)	0 (0.0)	1 (16.7)	3 (50.0)

Of 252 patients, 223 [88.5% (95% CI: 83.9-92.2)] reported at least one episode, totalling 2,118 crisis episodes throughout the year. Among the majority of patients (n=182, 72.2%; 95% CI: 66.3-77.7), three or more episodes of VOC per year were reported. The number of episodes per 1000 patient-year was 8404 (95% CI: 8040-8760) when all patients were used as the denominator, and 9490 (95% CI: 9090-9890) when only patients reported at least one episode were included. These episodes lasted for a median (IQR) duration of 4 (3-6) days.

Most patients (n=213, 95.5%; 95% CI: 91.9-97.8) sought treatment at least once, covering 1031 [48.7% (95% CI: 46.5-50.8)] of episodes, from public facilities, private providers, or traditional healers, *etc.* Hydroxyurea was consumed during the preceding fortnight in most episodes (n=2061, 97.3%; 95% CI: 96.5-98.0) among 214 (96% 95% CI: 91.9-97.8) of patients; however, use was largely episodic rather than continuous. Among patients reporting VOCs, 4% (n=9) had never consumed, 51.6% (n=115) had consumed ≤ 10 times annually, and the remainder >10 times. Among those without VOCs, 27.6% (n=8) had not taken hydroxyurea, and others reported infrequent use. Overall, 83 (3.9%; 95% CI: 3.1-4.8) of episodes required hospitalisation among fewer than one-fifth of patients, while 13 (0.6%; 95% CI: 0.3-1.1) required blood transfusions. Hospitalisations were mainly due to severe pain requiring parenteral analgesia, febrile illness or suspected infection, severe anaemia necessitating blood transfusion, and other acute SCD-

related complications. **Table II** shows the age-group-wise incidence of VOCs throughout the year among males and females. In the 0 to 10 years and 11 to 20 years age groups, the highest number of patients was recorded, with 1 to 5 crisis episodes per year. Whereas, in the 21- to 30-yr age group, the proportions of male patients with 1 to 5, 11 to 15, and 16 to 20 episodes were similar.

Figure shows that the first fortnight visit reported the highest number of illness episodes and the highest number of patients across all visits

SCD-related signs and symptoms, across three domains: general signs and symptoms, stroke-related symptoms, and splenic sequestration-related symptoms, observed over a year, are presented in **Table III**. Among general signs and symptoms, pain anywhere in the body was recorded in almost all episodes among patients. This was followed by fever, reported in 82.1% of patients across 57% of episodes.

Among stroke-related symptoms, sudden severe headache was the most prevalent symptom. In splenic sequestration-related symptoms, stomach pain and sudden weakness were reported in more than half of the participants.

Discussion

This multi-centric community-based prospective cohort study was designed to document the epidemiology of VOCs, the most common clinical manifestation of

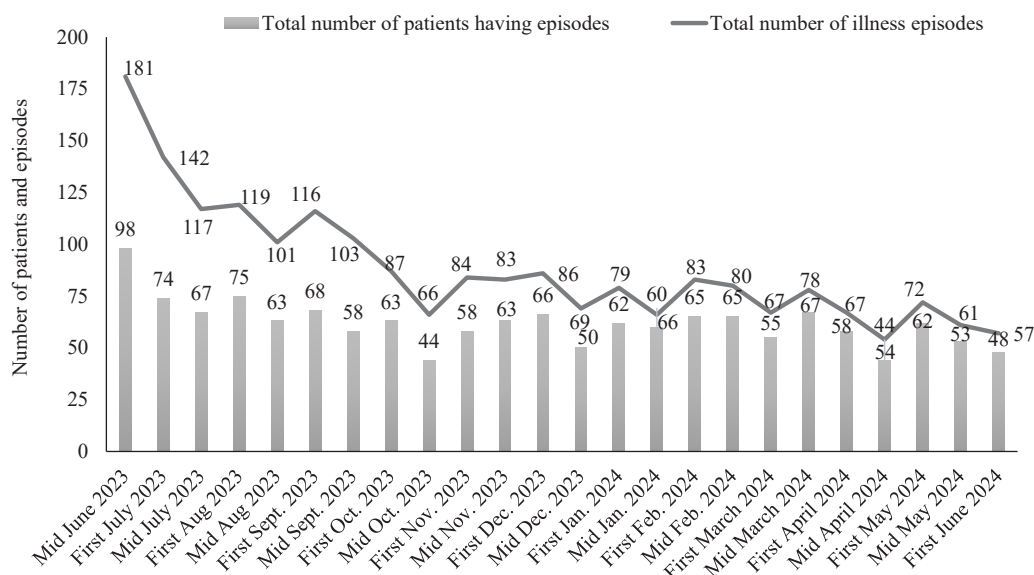


Figure. Fortnightly vaso-occlusive crisis episodes and the number of patients having them.

Table III. Signs and symptoms related to sickle cell disease observed in patients in a year

Variable	No. of patients; N=223, number (%; 95% CI)	Median (Q1-Q3) number of episodes per patient, N=223	No. of episodes; N=2118, number (%; 95% CI)	Median(Q1-Q3) duration per episode (in days), N=2118
General signs and symptoms				
Pain anywhere	217 (97.3; 94.2-99.0)	7.0 (3.0-13.0)	2042 (96.4; 95.5-97.2)	3.0 (2.0-5.0)
Swelling on joints	57 (25.6; 20.0-31.8)	2.0 (1.0-4.0)	227 (10.7; 9.4-12.1)	3.0 (2.0-5.0)
Pallor	48 (21.5; 16.3-27.5)	2.0 (1.0-4.0)	226 (10.7; 9.4-12.1)	4.0 (2.0-7.0)
Yellowness of eyes	67 (30.0; 24.1-36.5)	3.0 (1.0-5.0)	239 (11.3; 10.0-12.7)	3.0 (1.0-4.0)
Yellowness of skin	35 (15.7; 11.2-21.2)	1.0 (1.0-2.0)	58 (2.7; 2.1-3.5)	7.0 (4.0-9.0)
Swelling of abdomen	35 (15.7; 11.2-21.2)	3.0 (1.0-4.0)	135 (6.4; 5.4-7.5)	2.0 (1.0-4.0)
Breathlessness	70 (31.4; 25.4-37.9)	2.0 (1.0-4.0)	228 (10.8; 9.5-12.2)	2.0 (2.0-3.0)
Fever	183 (82.1; 76.4-86.9)	4.0 (2.0-9.0)	1207 (57.0; 54.9-59.1)	3.0 (2.0-4.0)
Stroke-related symptoms				
Sudden severe headache	104 (46.6; 40.0-53.4)	4.0 (2.0-10.7)	761 (35.9; 33.9-38.0)	2.0 (2.0-3.0)
Sudden trouble in seeing	26 (11.7; 7.8-16.6)	1.0 (1.0-3.0)	48 (2.3; 1.7-3.0)	2.0 (1.0-3.0)
Dizziness	76 (34.1; 27.9-40.7)	2.5 (2.0-7.5)	405 (19.1; 17.5-20.9)	2.0 (1.0-3.0)
Sudden confusion	19 (8.5; 5.2-13.0)	3.0 (1.0-6.0)	105 (4.9; 4.1-6.0)	3.0 (2.0-3.0)
Sudden trouble in walking	59 (26.5; 20.8-32.8)	3.0 (1.0-5.0)	282 (13.3; 11.9-14.8)	3.0 (2.0-3.0)
Sudden numbness	43 (19.3; 14.3-25.1)	3.0 (2.0-5.0)	257 (12.1; 10.8-13.6)	2.0 (1.0-4.0)
Loss of balance	37 (16.6; 12.0-22.1)	3.0 (1.5-5.5)	204 (9.6; 8.4-11.0)	2.0 (2.0-3.0)
Lack of coordination	22 (9.9; 6.3-14.6)	3.0 (1.7-5.0)	92 (4.3; 3.5-5.3)	2.0 (1.0-3.0)
Splenic sequestration-related symptoms				
Stomach pain	130 (58.3; 51.5-64.8)	3.0 (1.0-7.0)	635 (30.0; 28.0-32.0)	2.0 (2.0-3.0)
Pale lips	34 (15.3; 10.8-20.7)	1.0 (1.0-3.2)	86 (4.1; 3.3-5.0)	4.0 (2.0-5.0)
Sudden weakness	129 (57.9; 51.1-64.4)	3.0 (2.0-9.5)	1001 (47.3; 45.1-49.4)	3.0 (2.0-5.0)
Rapid breathing	39 (17.5; 12.7-23.1)	3.0 (1.0-5.0)	162 (7.7; 6.6-8.9)	2.0 (1.0-4.0)
Excessive thirst	66 (29.6; 23.7-36.1)	3.0 (1.0-4.5)	304 (14.4; 12.9-15.9)	2.0 (2.0-4.0)
Rapid heartbeat	53 (23.8; 18.3-29.9)	2.0 (1.0-4.0)	216 (10.2; 8.9-11.6)	2.0 (1.2-3.0)
Abdominal tenderness	17 (7.6; 4.5-11.9)	2.0 (1.0-3.0)	55 (2.6; 2.0-3.4)	4.0 (3.0-6.0)
Easy bleeding	2 (0.9; 0.1-3.2)	--	4 (0.2; 0.1-0.5)	--

CI, confidence intervals; IQR, interquartile range; Q, quartile

SCD. The distinct features of this study were a year-long, community-based, fortnightly data collection to minimise recall bias and to capture episodes irrespective of management. We report an incidence of at least one VOC of 88.5% over one year. In contrast, a multi-centric study conducted by Seth *et al*²¹ over seven months in India reported a prevalence of at least one VOC episode of 33.5%, which is less than half the prevalence reported in our study.²¹ The difference in the findings of these two studies can be attributed to their study designs. In hospital-based studies, only episodes in which patients visited healthcare centres for care were reported, thereby underestimating the number of VOC episodes. Evidence also suggests that 51% to 79% of SCD patients manage their VOCs at home.³⁰ Other studies conducted in adolescents and adult SCD patients reported VOC prevalence of 53% to 75%,³¹⁻³³ whereas studies by Singh *et al*³⁴ and Madhavi *et al*³⁵ in patients aged 6 months to 12 years reported VOC prevalence of 36.7% and 60%, respectively.

Another striking finding of our study was that the majority of patients received treatment at a healthcare facility for at least one VOC; however, the number of episodes treated at a healthcare facility was less than half, indicating a higher proportion of episodes were self-managed at home. It is common practice in these tribal settings to self-manage VOCs at home; if symptoms do not improve, individuals then visit healthcare facilities at a later stage. This behaviour has a bearing not only on the high morbidity but also on the mortality. Our study reported hospitalisation and blood transfusion rates of 19.7% and 5.4%, respectively. In contrast, a few hospital-based studies reported these events up to 89% and 77%.^{36,37} Lower rates of hospitalisation and blood transfusion in our study, despite the higher prevalence of stroke-related symptoms, can be attributed to limited access to tertiary healthcare facilities due to geographically isolated tribal habitations. Further, clinicians' prescriptions and patient compliance with hydroxyurea are suboptimal in India, despite its proven benefits.³⁸ However, 96% of patients consumed hydroxyurea during most VOCs in our study.

The five most prevalent signs and symptoms reported by our study participants were pain anywhere in the body, fever, abdominal pain, sudden weakness, and sudden severe headache. Other Indian studies have also reported pain as the most common symptom with varying frequency, ranging from 30% to 87%.^{31-34,39} The prevalence of various stroke-related symptoms in our study is much higher than the prevalence reported

in other studies.^{34,35} However, all of these studies were clinic-based and cross-sectional.

The findings of this study have direct relevance for the NSCAEM, which aims to strengthen early diagnosis, linkage to care, and long-term management of individuals with SCD through the public health system. The high burden of VOCs observed in this cohort highlights the need for consistent follow up, timely access to acute pain management, and improved continuity of care. Strengthening availability and adherence to hydroxyurea therapy, along with counselling, should be prioritised within the NSCAEM's implementation framework. Incorporating routine monitoring of VOCs and healthcare utilisation into programmatic reporting may help identify high-risk individuals and guide targeted interventions in resource-limited tribal settings.

This study provides strong evidence on the epidemiology of VOCs, owing to its robust study design, higher follow up rates, and other distinct features discussed above. However, the study has a few limitations. The study was conducted in purposively selected SCD-endemic tribal districts using purposive sampling to include all available and consenting registry patients. This may limit the generalisability of the findings beyond similar programmatic and tribal settings, and some degree of selection bias cannot be ruled out. Consequently, the findings may not be directly applicable to all SCD populations in India. While the study draws on data from geographically distinct settings, unmeasured factors such as socio-economic conditions, healthcare access, and programme implementation may have influenced the observed patterns. Some of the information, based on participants' self-reports, may be subject to reporting and recall bias. Although we attempted to corroborate self-reported data with medical records, where available, these records were not accessible for cases managed by private pharmacies and unqualified practitioners. However, the data were collected by trained research personnel, which minimises the risk of discrepancies. Despite these limitations, the study provides important insights into the burden and patterns of VOCs.

The study findings play a crucial role in demystifying the notion that SCD is milder and less severe in India. This first-ever community-based study highlights that the burden of morbidity among SCD patients in India is much higher than estimations provided by hospital-based studies. Our study lays the groundwork for future longitudinal assessments of clinical outcomes. It also

शोध-संदेश

भारत में सिकल सेल रोग (SCD) को लंबे समय से अपेक्षाकृत हल्का रोग माना जाता रहा है, किंतु देशव्यापी ठोस आंकड़ों के अभाव में इसके वास्तविक रोग-भार का आकलन सीमित रहा है। वेसो-ऑक्लूसिव क्राइसिस (VOCs) इस रोग की सबसे सामान्य और कष्टदायक जटिलताओं में से एक है, जो रोगियों के जीवन-स्तर को प्रभावित करती है। प्रस्तुत सामुदायिक-आधारित बहु-केंद्रीय अध्ययन में अधिकांश रोगियों में संकट की घटनाओं की अधिक संख्या पाई गई है, जो भारत में SCD के कम गंभीर होने की धारणा को चुनौती देती है। साथ ही, स्वास्थ्य सेवाओं के कम उपयोग की प्रवृत्ति भी सामने आई है, जो चिंता का विषय है। यह अध्ययन SCD के प्रभावी प्रबंधन, बेहतर निगरानी तथा स्वास्थ्य सेवाओं को सुदृढ़ करने की आवश्यकता को रेखांकित करता है।

highlights the higher proportion of VOC episodes that are self-managed at home. Future research should focus on evaluating programme implementation, identifying context-specific barriers to hydroxyurea adherence and timely access to acute care, and assessing scalable primary healthcare-based models to reduce SCD-related morbidity. In addition, studies examining the role of genetic modifiers, including HbF levels and other haemoglobin variants, in influencing the frequency and severity of VOCs would help improve risk stratification and guide individualised approaches to care.

Our study cohort, followed for one year, exhibited a higher prevalence of VOCs. The management of VOCs indicates inappropriate and inconsistent consumption of hydroxyurea. Lower rates of hospitalisation and blood transfusion indicate that tertiary-level healthcare in tribal areas needs improvement. Enhanced awareness, counselling, and optimisation of hydroxyurea use may contribute to improved management of VOCs in underserved settings. The findings of this study will undoubtedly aid in taking definitive actions to reduce the burden of SCD-related morbidity.

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