J Nepal Health Res Counc 2025 April-June; 23 (67): 361-368

Socio-Demographic Characteristics and Clinical profile in Sickle Cell Disease Patients

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ABSTRACT

Background: Sickle cell disease (SCD) is a significant public health issue in Nepal, predominantly affecting the Tharu community in the Mid-Western and Sudurpaschim province. This study was design to understand the socio-demographic characteristics and symptoms of SCD patients attending the health camp conducted by Seti Provincial Hospital in Sudurpashchim Province, Nepal.

Methods: This study was cross-sectional study conducted at Sudurpashchim Province of Nepal visiting in Seti provincial Hospital during the period of free health camp organized by Hospital.

After ethical clearance, patients were enrolled based on inclusion and exclusion criteria, and their demographic, epidemiological, and clinical profiles were recorded using a structured questionnaire.

Results: Among 119 patients with SCD, the mean age was 22.58 years, with a majority (58.8%) being female. Most patients (45.37%) belonged to the upper lower socioeconomic class. The most common symptom was musculoskeletal pain, weakness, and fever (30.5%), followed by jaundice, chest pain, and vertigo (21.84%). Additionally, 15.96% experienced weakness, nausea, breathing difficulties, and fever; 12.60% reported weight loss, low hemoglobin, and chest pain; 10.08% had backache, joint pain, fever, and headache; and 9.24% experienced knee pain, fever, anemia, and vertigo.

Conclusions: SCD is seen in younger patients in Sudurpashchim Province, especially Kailali and Kanchanpur Districts with Tharu communities are more affected. Major systemic manifestations of SCD include pain crisis, hemolytic crisis, acute chest syndrome, hepatopathy and AVN of hip. The government of Nepal should implement policies focus for treatment and pain management within the current health system, to minimize, control, and prevent the high burden of SCD in the Tharu communities.

Keywords: Haemolytic anemia; Nepal; pain crises; sickle cell disease.

INTRODUCTION

SCD is known as genetic blood disorder diseases one of significant public health issue in Nepal, predominantly affecting the Tharu community in the Mid-Western and Sudurpaschim province.[1-3] It is a genetic disorder that causes hemoglobin to become rigid and sickle-shaped, disrupting oxygen transport. SCD is prevalent in sub-Saharan Africa, the Middle East, India, the Mediterranean, and Nepal, but in these regions remains underexplored.[4-6] SCD begins at 5-6 months, causing pain, anemia, and swelling. SCD occurs when a person inherits two abnormal ß-globin genes, one from each parent, affecting hemoglobin that managed with

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hydration, a healthy diet, pain relief, hydroxyurea, and gene therapy.[7-12] SCD is common in the Tharu and Rana Tharu communities in Sudurpashchim Province, Nepal, where urbanization and migration are increasing.[11-13] However, research is limited and identifying risk groups could help reduce morbidity and support policy development. This study aims to understand the socio-demographic characteristics and symptoms of SCD patients attending health camps.

METHODS

This was a descriptive cross sectional study conducted in Kailali and Kanchanpur districts of Sudurpashchim province of Nepal with covers 19,515.52 km^2 area (Figure 1).

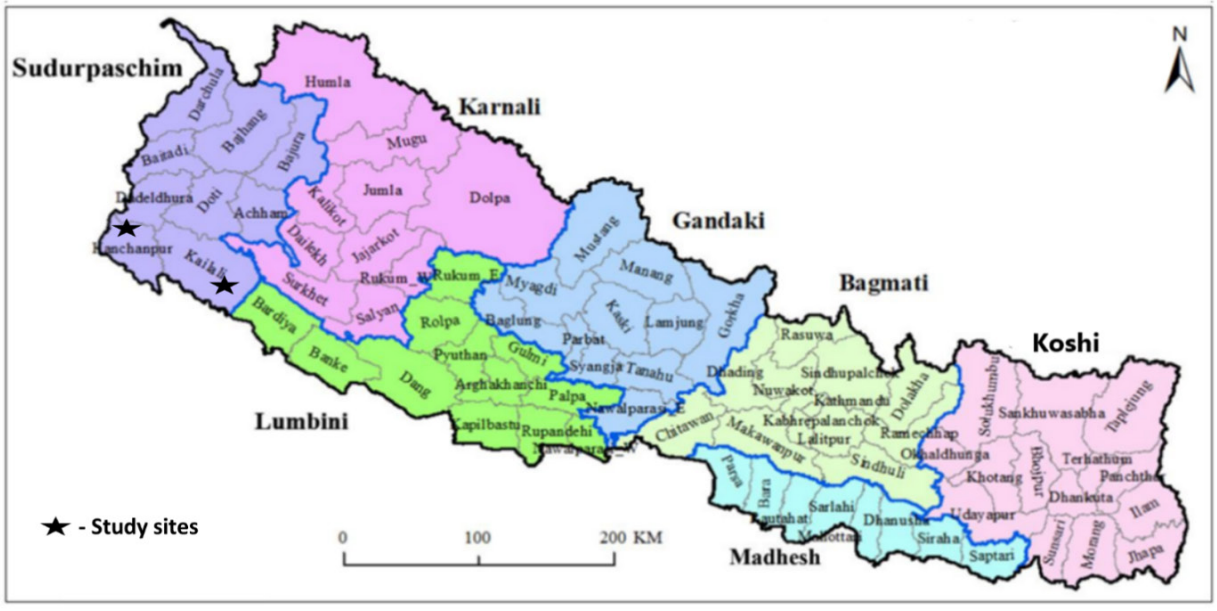

Figure1. Map of Nepal showing study sites.

A registered sickle cell disease patient in Seti Provincial hospital who are already diagnosed and have been on continuous treatment was the study group. All interested sickle cell disease patients attending the health camp conducted by Seti Provincial Hospital and willing to participant with age above 18 years were included in this study. While those who do not willing to participate in this study and under age of 18 year were excluded from the current study.

Before conducting the research, approval was taken from concerned authority of Seti Provincial Hospital, Dhangadhi and Nepal Health Research Council, Kathmandu (Ref No:3170/2021). Written informed consent was taken from each respondent before taking the interview.The Government of Nepal provides free health services through the "Bipanna Nagarik Upchar Kosh Program" at Seti Provincial Hospital. The study will include patients over 18 who consent to participate, with the sample size based on research by Marchand M et al,[13] which found a 9.3% prevalence of Hbs in the Tharu population of Dang, Nepal.

In this study, the calculated sample size was 129.61 based on the formula, but only 119 eligible samples were taken.

The cross-sectional survey with convenience sampling was done and altogether 119 patients with SCD conditions were enrolled in this study. Structured questionnaire was introduced by interviewer to each sickle cell disease patients. An informed consent and willingness of participant were taken before proceeding for interview. Socio demographic, dietary and health seeking data including age, sex, address, religion, education, ethnicity, income

status, dietary pattern, addiction of alcohol and smoking, patient's medication like hydroxyurea and opioid, meditation, social activity, height and weight were recorded by interview. For the measurement of weight, weighing machine was used and inch tap was used for height measurement. After collection of data, they were verified and entered into Micro soft Excel and analyses were performed with the data analysis by Microsoft Excel.

RESULTS

The majority of respondents (52.1%) were aged 19-29 years, followed by 26.89% in the 29-39 year age group. Interestingly, the longest surviving patients with SCD were found to be in the 59-69 age group. Of the total respondents, 58.8% were female and 41.2% were male. Most respondents (56.3%) were Tharu, followed by Chaudhary (31.9%) and Rana (11%). A majority (86.6%) were Hindu, while 12.6% were Christian. Most respondents (82.35%) lived in Kailali district, with 17.64% residing in Kanchanpur. Nearly all respondents (99.2%) were local residents, while 0.8% was migrants from hilly areas in Table1.

The house of 82.4% of respondents were kachi (Temporary house-temporary or makeshift houses made of non-durable materials like mud, thatch, bamboo, or other natural materials), 6.7% lived in semi pucca (Semi-permanent houses made of a combination of durable and non-durable materials, such as brick walls with a thatched or tin roof) and only 10.9% had pucca house (ppermanent houses constructed entirely of durable materials like brick, cement, concrete, or steel (Figure 2).

The educational status of most of respondents were secondary level (37%) followed by primary level (28.6%) and graduate (6.7%) while 26.9% were literate. Respondents were involved in different work with highest percentage accounted by farm work (42.9%) followed by labour (35.3%), business (10.9%), service (10.1%). Similarly almost half (45.37%) of respondents belonged to upper lower class, 32.77 % to lower class, 15.12 % to lower middle class, 4.20 % to upper middle class and only 2.52 % were upper class (Table 2).

The majority of the respondents (83.2%) were non vegetarian, whereas only 16.8 % were vegetarian. The junk food consumption were high (62.2%) among the respondents. Taking about addiction, 40.33 % of respondent had no history of addiction while 18.80 % had addiction of alcohol and smoking, 15.96 % only addicted to alcohol, 15.12 % addicted for tobacco chewing, 6.71 % had habit of smoking and 5.04% were addicted to all three substance Tobacco chewing, smoking and alcohol (Table 3).

The most common signs and symptoms of respondents (30.5%) were weakness, joint pain, and headache. Body ache, fever on off, 21.84 % had symptom of chest pain, jaundice, vertigo, joint pain, 15.96 % felt weakness, nausea, difficulty in breathing, fever, 12.60% experienced weight loss , loss of hemoglobin, chest pain, 10.08% explained backache, multiple joint pain, fever, headache and 9.24 p had Pain in knee joint, fever, anemia, vertigo (Table 4).

The BMI of 70.6% of respondents was normal, 24.4% were underweight, and 0.8% were obese. Hydroxyurea, folic acid, and ketrol were commonly used by 86.55% of respondents. Folic acid alone was consumed by 95.79%, while 55.46% took both folic acid and ketrol, and 12.60% used other painkillers. About half of the respondents (47.05%) visited their physicians every more than 2 months, 40.7% visited every 2 months, and 12.6% visited monthly. During visits, 31.93% spent 100-500 NRs, 22.68% spent 500-1500 NRs, 19.32% spent 1500-2500 NRs, 14.28% spent 2500-3500 NRs, and 10.08% spent 3500-4500 NRs. Most respondents (78.99%) regularly consulted physicians for biomedical tests and medications, 65.54% for multi-organ issues, and 40.33% for joint pain and blood transfusions. A high number of respondents' family members (84.87%) had SCD (Table 5).

Among the diagnosed patients, 79% reported no tension about their disease whereas 21% respondents experienced stress related to their disease condition. To over the stress, 31.93% were participating daily in religious activities and 21.8% participating in weekly basis. Likewise, 61.34% respondents were involved in physical exercise on daily basis, 4.20 % performed on weekly basis. Moreover, 65.54 % respondents had reported to undergo the training regarding management of their disease (Table 6).

Among the respondents, 93.27% received organizational support for their treatment, while 6.72% did not. Additionally, the majority (65.54%) expected free checkups, lifetime medication, and treatment. Meanwhile, 19.32% requested the availability of physicians in government hospitals, and 15.12% wanted SDC to be included as a priority program in the health system and research (Table 7).

Table 1. Socio-demographic profile of respondents (n=119)

Socio-demographic variable	Frequency (N)	Percentage (%)
Gender		
Male	49	41.2
Female	70	58.8
Total	**119**	**100.0**
Age group		
19- 29	62	52.1
29-39	32	26.89
39-49	12	10.8
49-59	12	10.8
59-69	1	0.8
Cast of respondent		
Tharu	67	56.3
Chaudary	41	35.5
Rana	11	9.2
Religion		
Hindu	103	86.6
Christian	15	12.6
Other	1	0.8
Living District of Respondents		
Kailali	98	82.35
Kanchanpur	21	17.64
Residence		
Local	118	99.2
Migrated	1	0.8
Family structure		
Nuclear Joint	73 46	61.3 38.7
Marital status		
Married	89	74.8
Unmarried	28	23.5
Divorced	2	1.7
Type of house		
kachi	98	82.4
Pucca	13	10.9
Semi pucca	8	6.7

Table 2: Educational and economic background of the respondents (n=119)

Variable	Frequency (N)	Percentage (%)
Education		
Illiterate	32	26.9
Primary	34	28.6
Secondary	44	37
Graduate9	8	6.7
PG and above	1	0.8
Occupation		
Service	12	10.1
Business	13	10.9
Farm work	51	42.9
Labour	42	35,3
Pension	1	0.8
Economic Grade cccording to the Kuppuswami's classification		
Upper	3	2.52
Upper middle	5	4.20
lower middle	18	15.12
Upper lower	54	45.37
Lower	39	32.77

Table3: Dietary pattern and food habit of the respondents (n=119)

Characteristics	Frequency (N)	Percentage (%)
Dietary habit		
Non vegetarian.	99	83.2
Vegetarian	20	16.8
Consumption of junk food		
Yes	74	62.2
No	45	37.8
Addiction of respondents		
Tobacco(chewing)	18	15.12
Smoking	8	6.71
Alcohol	19	15.96
Alcohol and smoking	20	16.80
Tobacco chewing , smoking and alcohol	6	5.04
No addiction	48	40.33

Table 4. Sign and symptoms during first visit to doctor ((n=119)

Sign and symptoms	Frequency (N)	Percentage (%)
Felling weakness, joint pain, and headache. Bodyache, fever on off	36	30.25
Chest pain, Jundic, vertigo, joint pain	26	21.84
Weakness, nausea, Difficulty in breathing, fever	19	15.96
Weight loss , loss of hemoglobin, chest pain	15	12.60
Pain in knee joint, fever, anaemia, vertigo	11	9.24
Backache, Multiple joint pain, fever, headache	12	10.08
Total	**119**	**100.0**

Table 5. Health seeking of the respondents (n=119)

Characteristics	Frequency (N)	Percentage (%)
BMI		
Under weight (< 18)	29	24.4
Normal(18 -24.99)	84	70.6
Over weight (25-29.99)	5	4.2
Obese (> 30)	1	0.8
Medicine taken		
Hydroxyurea , folic acid, ketrol	103	86.55
Folic acid only	114	95.79
Folic acid , Pain Killer (Ketrol)	66	55.46
Other Pain killer	15	12.60
Duration of visit to health worker and physician		
Every month	15	12.6
Every 2 month	48	40.3
More than 2 month	56	47.05
Expenses (NRs) for every visit to health worker and physician		
100 -500	**27**	**22.68**
500-1500	38	31.93

Table 5. Health seeking of the respondents (n=119)

Characteristics	Frequency (N)	Percentage (%)
1500-2500	23	19.32
2500-3500	17	14.28
3500-4500	12	10.08
4500-5500	1	0.8
> then 5500	1	0.8
Consultation for health problems during regular check up	**frequency**	**percentage**
Multi organ problem with Biomedical test	78	65.54
Multiple Joint pain and Blood transfusion	48	40.33
Biomedical test and medicine	94	78.99
Family Member having SCD		
Yes	101	84.87
Unknown	18	15.12

Table 6: Stress after diagnosis SCD and their management (n=119)

Characteristics	Frequency (N)	Percentage (%)
Stress after diagnosis SCD		
Yes	25	21
No	94	79
Religious activity		
Never	20	16.80
Daily	38	31.93
Weekly	26	21.84
only festival	35	29.41
Exercise schedule		
Daily	73	61.34
Weekly	5	4.20
Monthly	1	0.8
No exercise	40	33.91
Taken SDC management training		
Yes	78	65.54
No	41	34.45

Table 7: Support and expectation from different organization and government (n=119)

Characteristics	Frequency (N)	Percentage (%)
Support from organization		
Yes	111	93.27
No	8	6.72
Expectation form government		
Free checkup, life time medicine and treatment	78	65.54
Availability of physician in government hospital	23	19.32
Research and Include as priority program in health system	18	15.12

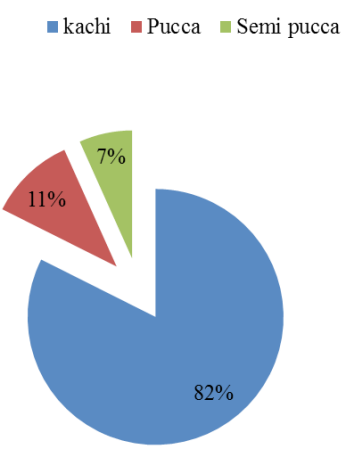

Figure 2. Respondents by type of house. (n=119)

DISCUSSION

The first case of SCD was recorded and reported in 2003.[14] Since, then the government of Nepal has decided to fund SCD treatment through the dis-advantaged citizens Fund.[12] However, receiving financial assistance for SCD therapy is contingent upon receiving a diagnosis, thus it is critical to have a healthcare infrastructure in place to facilitate this process. At the same time most patients with SCD and other haemoglobinopathies in Nepal are getting diagnosed later in their lives which made difficult to manage the condition. For instances, majority of respondents in this study was in the age group 19-29 years, followed by age group 29-39 years. Similar pattern was observed in previous study from Nepal.[2] Thus, some previous studies revealed that lower age group i.e. 11-20 years are affected most.[15,16]

Previous studies indicate that the majority of respondents were female, consistent with a similar study conducted in Nepal.[2] This finding contrasts with other studies.[16,17] The current study noticed that more than half of study participants only had primary or secondary level education which definitely hampers their heath seeking behavior as well as preventive measure. Majority of patients had family history of sickle cell disease. This means diseases was caused by genetic disorder and was transmitted from generation to generation.

This investigation demonstrates that SCD are widespread among Tharu ethnic group. So far, the majority of those afflicted are still undiagnosed because there are not any specialized laboratories testing available in that area.[13] The majority of patients were local residences whereas some were migrated Indian origin Rana Tharu. This showed the chance of inheriting sickle traits from India which the highly endemic country for SCD. The disease primarily affects scheduled castes, scheduled tribes, and other lower social groupings all over the world.[18,19] Neonatal screening is strongly advised in order to identify and treat children with SCD; affordable interventions, such as vaccinations and penicillin prophylaxis, can lower the risk of infection in infancy and potentially extend the life expectancy of SCD patients.[20,21]

This investigation shows that SCD is widespread among the Tharu ethnic group. However, many affected individuals remain undiagnosed due to the lack of specialized laboratory testing in the area.[13] Most patients are local residents, with some being migrant Indian-origin Rana Tharu, suggesting a potential inheritance of sickle traits from India, a country highly endemic for SCD. The disease primarily affects scheduled castes, scheduled tribes, and other lower social groups worldwide.[18,19] Neonatal screening is strongly recommended to identify and treat children with SCD; affordable interventions such as vaccinations and penicillin prophylaxis can reduce the risk of infection in infancy and potentially extend the life expectancy of SCD patients.[20,21]

Two third of the patients belonged to the lower class sustaining their life in the agricultural works. At a same time, study pointed that patients had to expense NRs 500 -5500 for every visit to health care facility which is very difficult for the people under poverty line. It is true that most of the patients getting support from government for the treatment whereas some patients does not getting any support at all and health care is

difficult to them.

Study revealed the dietary habit of most of the respondents (83.2%) were non vegetarian who also consumed the junk food. This suggests those respondents had unhealthy food habit which may be responsible for comorbid of other disease condition. Study revealed that more than half patients had addiction of alcohol or smoking or tobacco or all. This type of addiction might be the result of stress full condition, which further contributes to worsen the health condition. There was significant relation between addiction and severity of disease.

Health-seeking behavior is among the important factors determining uptake and outcomes of health care services for sickle cell disease. Health-seeking behavior is influenced by peoples' perceptions about the disease within the context of tradition, cultural beliefs and attitudes. Most of the respondents with sickle cell disease reported experiencing a number of signs and symptoms such as joint pain, jaundice, anemia, fatigue, body weakness, ongoing anxiety and low sexual dysfunction. These signs and symptoms and their severity led them to seek care and manner in which they manifested themselves alerted patients and their family members or social networks that something was not right and that they needed to go somewhere to get some explanations.

Majority of respondents were participating in religious activities and physical exercise daily or weekly basis. Moreover 54% respondents had reported to undergo the training regarding management of their disease. This is the good aspects for the management of the disease. For the control and prevention of the complication of sickle cell disease counseling after care and health seeking behavior has to be enhanced by the physician. This part is neglected by the attending physician to their patients in context of Nepal which needs to be rectified.

CONCLUSIONS

SCD is prevalent in the Kailali and Kanchanpur districts among the Tharu community in the Sudurpashchim Province of Nepal. Public awareness about SCD and health-seeking behavior is lacking, posing a significant public health challenge and an alarming threat to the national health system for controlling and preventing SCD. The disease primarily affects the upper lower and lower classes, particularly those working on farms. A majority of respondents had a history of smoking, chewing tobacco, and alcohol addiction. Common symptoms included weakness, joint pain, headache, jaundice, body ache, and fever. Patients with SCD incur expenses of 1000 NRs (ranging from 500 to 1500 NRs) per visit for medical consultation and biomedical tests, which they must undergo frequently. This highlights the need for the government to develop and implement a new strategic plan with pain management in the current health system building blocks, active community participation, to minimize, control, and prevent the high burden of SCD in the Tharu community.

ACKNOWLEDGEMENTS

The authors would like to extend their heartfelt gratitude to the staff at Seti Provincial Hospital, Ministry of Social Development Sudurpashchim Province, Dhangadhi, Kailali, Nepal, and all those who provided direct and indirect support. Additionally, we deeply appreciate the contributions of the study participants, whose involvement was crucial to the completion of this research.

CONFLICT OF INTERESTS

The authors declare no conflict of interests.

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