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RESEARCH

Comparative assessment of quality of life among adolescents with sickle cell disease and sickle cell trait: evidence from Odisha, India

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Abstract

The present study aims to assess the quality of life (QOL) of adolescents with sickle cell disease (SCD) and sickle cell trait (SCT) in hard-to-reach regions in Koraput district of Odisha state. 387 adolescents with sickle cell genes (HbSS = 52, HbAS = 135, HbA = 200) were selected through their medical records from southern parts of Odisha. A validated and pre-tested QOL scale was modified to assess the QOL. The questionnaires were modified by aiming to describe the proportion of adolescents who feel restricted in different domains, measuring the extent within each domain, and finding an aggregate score of QOL. Furthermore, to explore the expenditure on health, 552 households were selected randomly, of which 72 families had HbS individuals. This study found a significantly lower health-related QOL in adolescents with SCD. However, most psychosocial sub-domains, for instance, worry about the illness, frequency of angry days, feeling jealousness toward other normal adolescents, and negative feelings of sadness on some days, are similarly affected in adolescents with SCT and SCD. The overall QOL of SCD individuals is more affected (percentage of affected mean score = 60.93%), followed by SCT individuals (35.63%). Healthy adolescents' QOL is relatively unaffected (13% were affected). The yearly frequency of blood transfusion received (1.7 ± 0.4) and hospitalization (2.1 ± 0.9) was significantly higher in adolescents with SCD. The healthcare expenditure was significantly higher (3.6% to 81.3% of the family income) in families with HbS than in families without HbS (0.8% to 19.2%) ($p < 0.05$). The overall QOL was affected in both SCD and SCT adolescents. The focus should be given equally to both SCD and SCT individuals, in spite of only SCD individuals.

Keywords Sickle cell disease · Sickle cell trait · Quality of life (QOL) · Adolescents

Introduction

Sickle cell hemoglobinopathy includes all the abnormal hemoglobin S (HbS), i.e., sickle cell trait (HbAS), sickle cell disease (HbSS), and an array of miscellaneous heterozygous hemoglobinopathies such as hemoglobin SC disease, hemoglobin SD disease, hemoglobin SO Arab disease, and HbS combined with beta-thalassemia (Hb S/β Th). Around 300 million sickle cell carrier individuals are present worldwide, with concentrations in Africa, the Arab Peninsula, India, the Mediterranean, and the southern United States (Key et al. 2015). The sickle gene is prevalent in numerous tribal communities in India, with a prevalence of 1 to 40% (Colah et al. 2015), who have a high frequency of socioeconomic disadvantage and are medically underserved (Bindhani et al. 2020). SCD has high degrees of comorbidities and mortality, and QOL among adolescents having SCD has not been broadly reported. Furthermore, very few studies have been reported concerning the QOL among adolescents with SCT, which is relatively asymptomatic and central to the present study.

Though it is well established that Odisha is at higher risk for sickle cell hemoglobinopathies, there is a lack of an appropriate approach to initiate a large-scale population-based study that covers all the sickle cell prevalent areas (Bindhani et al. 2020; Mohanty and Das 2011). Koraput district of Odisha state, where the present study was conducted, is one of the underdeveloped districts and predominantly inhabited by tribal and scheduled caste people, who have a high dominance of socioeconomic disadvantage and are often medically underprivileged (Bindhani and

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Nayak 2018). There is no published in-depth population-based study assessing their quality of life. Also, not a single study was found that focuses on adolescents with sickle cell hemoglobin.

Thus, the present study aims to assess the QOL by exploring specific domains that are most affected in adolescents with sickle cell disease (SCD) and sickle cell trait (SCT) in hard-to-reach regions in Koraput district of Odisha.

Materials and methods

The present retrospective cross-sectional study was carried out in Koraput district of Odisha, India, from December 2020 to July 2023, spanning more than 2.5 years. The snowball sampling technique was used with the help of villagers, Anganwadi and health personnel to find villages having a high number of individuals with sickle cell genes. Villages ($n=26$), that were remotely situated and where healthcare facilities were not easily accessible, were finally picked up for this study. From these villages, a total of 387 adolescents of both sexes in the age group of 10–17 years were selected from their medical/caregiver records. Out of the 387 recruited samples, 52 had sickle cell disease (SCD), and 135 had SCT. Additionally, 200 age-sex-community-matched normal controls were interviewed to compare their QOL to the adolescents with SCD and SCT.

To evaluate QOL, the present QOL scale was modified from the model developed by Patel and Pathan, which developed based on the multidimensional conceptual framework of QOL by Patrick and Erickson, EUROQOL, and WHO QOL BREF (Patel and Pathan 2005). This questionnaire was specifically designed to allow children aged 8 years and older to articulate the impact of illness on their quality of life across different areas or domains. The present scale assessed physical function, psychological function, social function, health perception, morbidity, and opportunity, each further divided into various sub-domains as detailed in the results section. Further, this scale was empirically validated after a pilot study. This study aimed to describe the proportion of adolescents who feel restricted in different domains and measure the extent within each domain or have an aggregate score of quality of life. Therefore, this validated scale could help to enhance the quality of the study. Adolescents in the age group of 10–17 years old are better at responding to the questionnaire. Additionally, necessary help, when required, was taken from the parents of the subjects during the data collection. The demographic data and medical history of the participants were collected using a scheduled method. Moreover, to explore the expenditure on health, 552 households were selected randomly, of which 72 households had HbS individuals.

All statistical analyses were conducted using SPSS (version 22). Statistical significance was determined at a p-value of ≤ 0.05. Microsoft Excel was utilized for data sorting.

The study received approval from the Institutional Ethical Committee. Prior to recruitment, informed written consent, transcribed in local languages, was obtained from both participating adolescents and their parents.

Results

Of the 387 recruited adolescents, 173 (44.7%) were males and 214 (55.3%) were females. Further, 52 individuals were with SCD (HbSS), 135 were with SCT (HbAS), and 200 were without sickle cell hemoglobin or controls (HbA). The mean age (±SD) was found to be 15.2 ± 2.8. The frequency of blood transfusion received (1.7 ± 0.4) was significantly higher in adolescents with SCD than in SCT (0.07 ± 0.3) and healthy controls (0) during the last 1 year from the day of the survey. The notable factor here is that individuals with SCT were found to have undergone blood transfusions in some instances. Furthermore, the frequency of hospitalization (2.1 ± 0.9) was found to be significantly higher among adolescents having SCD (Table 1).

Figure 1 represents the overall QOL score of respondents with HbSS, HbAS and HbAA, respectively. The higher score represents a better quality of life. The QOL of HbSS individuals was found to be highly affected, followed by that of

Table 1 Comparison of sociodemographic and medical history information among individuals with SCD, SCT, and normal status ($n=387$)

Characteristics	SCD ($n=52$)	SCT ($n=135$)	Normal/HbA ($n=200$)	p-value
Male (No., %)	24 (46.15)	58 (42.96)	91 (45.5)	0.878
Female	28 (53.85)	77 (57.04)	109 (54.5)	
Age (mean ± SD) (in years)	14.7 ± 2.4	15.1 ± 2.6	15.8 ± 3.5	0.891
Educational level (mean ± SD)	5.2 ± 2.1	6.4 ± 2.4	6.7 ± 2.5	0.345
Annual family income (mean ± SD) (in INR)	62,035.1 ± 16,792.9	62,642.3 ± 15,163.8	63,173.9 ± 18,792.1	0.427
Medical history				
No. of times hospitalized (in last 1 year)	2.1 ± 0.9	0.8 ± 0.4	0.3 ± 0.3	0.03
Transfusion received (in last 1 year)	1.7 ± 0.4	0.07 ± 0.3	0	< 0.001

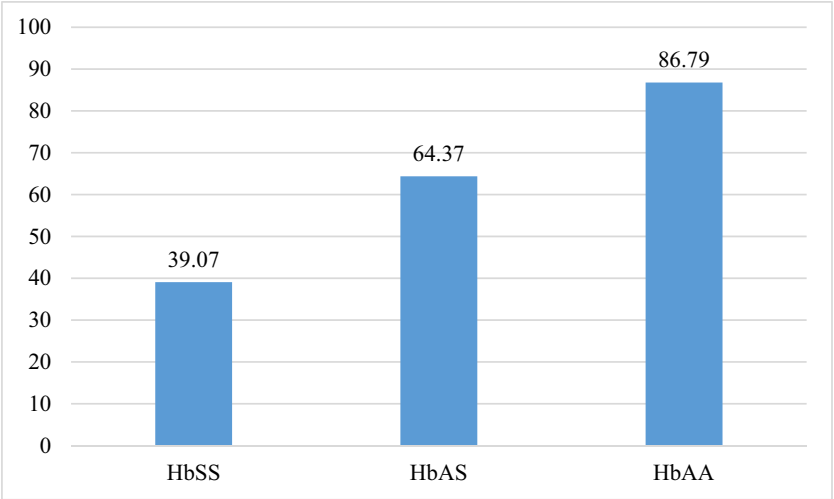

Fig. 1 Comparisons of overall QOL score of respondents with HbSS, HbAS and HbAA (p value < 0.05)

HbAS individuals. The QOL was better among the individuals without HbS. These scores have significantly differed among the three categories. Analysing the various domains (physical function, psychological function, social function, morbidities and opportunities) revealed that the QOL of all these domains were affected (Fig. 2). Further, the QOL in each domain significantly differed among the HbSS, HbAS, and HbAA individuals. Interestingly, the control group, consisting of HbA individuals, was categorized based on the presence of family members with HbS. It was found that the quality of life (QOL) of HbA individuals who have family members with HbS was significantly lower compared to HbA individuals without HbS family members ($p < 0.05$) (Fig. 3).

Tables 2, 3, 4, 5, 6, and 7 depict the differences in quality of life among adolescents with SCD, SCT and normal controls in various domains like physical, psychological, cognitive, and social functions, along with their health, comorbidities, and perceptions of opportunities. As expected, all the domains were relatively unaffected in normal adolescents compared to adolescents with sickle cell hemoglobin (HbS). Most of the domains (physical, psychological, cognitive, and social functions) were affected in adolescents with SCD, followed by SCT. However, most psychosocial sub-domains, for instance, worry about the illness, frequency of angry days, feeling jealousness to other normal adolescents, and negative feelings of sadness on some days, are similarly affected in adolescents with SCT and SCD. Further, the study reported that adolescents with both SCD and SCT were getting equal support from their teachers and friends. In the domain of health perception, it is noteworthy that

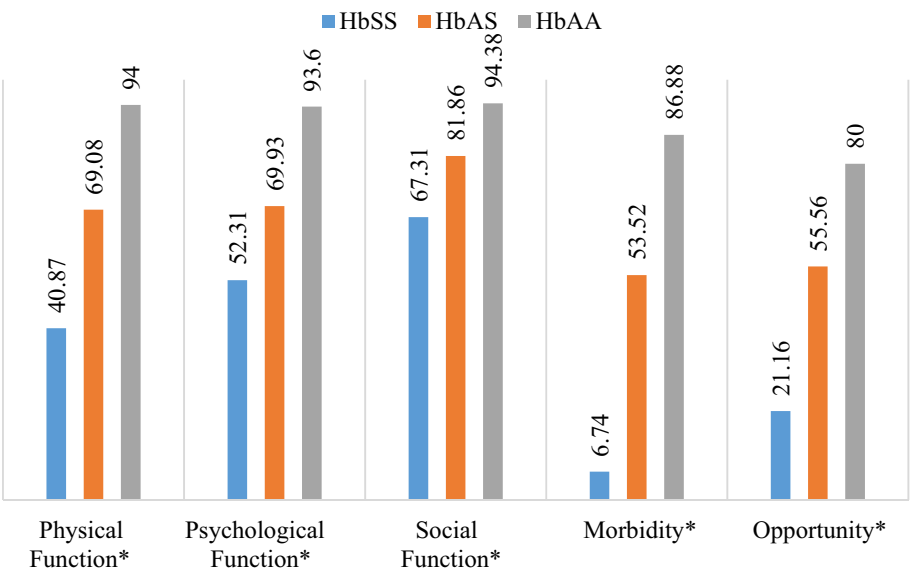

Fig. 2 Domain wise comparison of QOL (*$p < 0.05$)

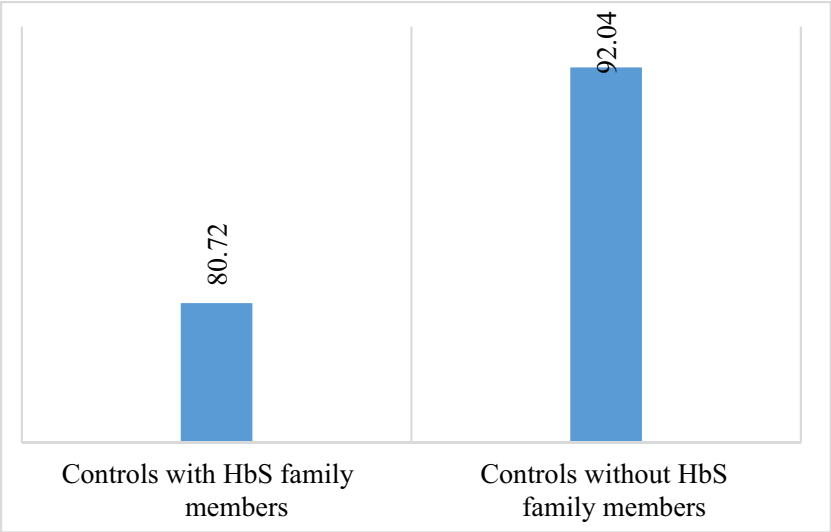

Fig. 3 Comparison of QOL score among normal individuals (HbA) with HbS family members and without family members ($p < 0.05$)

Table 2 Adolescents' responses for different items of physical function

Physical function	HbSS		HbAS		HbA		
	No	%	No	%	No	%	
Mobility							
No problem	36	69.23	124	91.85	191	95.50	< 0.001
Problem on some days	11	21.15	11	8.15	9	4.50	
Unable to do on most days	5	9.62	0	0.00	0	0.00	
Indoor games							
No problem	31	59.62	102	75.56	193	96.50	< 0.001
Problem on some days	14	26.92	33	24.44	7	3.50	
Unable to do on most days	7	13.46	0	0.00	0	0.00	
Attendance in school							
No problem	10	19.23	68	50.37	180	90.00	< 0.001
Problem on some days	27	51.92	45	33.33	19	9.50	
Unable to do on most days	15	28.85	22	16.30	1	0.50	
Outdoor games							
No problem	8	15.38	79	58.52	188	94.00	< 0.001
Problem on some days	23	44.23	50	37.04	11	5.50	
Unable to do on most days	21	40.38	6	4.44	1	0.50	

adolescents with SCT experienced the common cold more frequently (62.96%) compared to their counterparts with SCD, who had a lower incidence rate of about 32.69%. In the sub-domain of feeling sad, both the groups (SCD and SCT) felt that they are the economic burdens on their families because of extensive hospitalization and health care services, which is another root cause for their sadness and depression. The study found breathing problems and weakness among most respondents having HbS in the studied area. The intensity of pain was significantly more severe in individuals with SCD than in SCT. As expected, the QOL is notably more affected in individuals with SCD. Nevertheless, there's also a distinct impact on the overall QOL among those with SCT to some degree.

The annual healthcare cost in households with HbS was significantly higher (3.6% to 81.3% of the total family income) than in households without HbS (0.8% to 19.2%). The families with only sickle cell carriers had lower spending than families with sickle cell disease. 8.33% of the households having HbS had more than 40% healthcare cost, whereas no households without HbS had more than 40% healthcare cost (Table 8). The annual healthcare cost was significantly higher among the households with HbS individuals (5182.5 ± 1124) than the households without HbS (911.4 ± 315.8).

More importantly, the correlation analysis revealed a positive correlation between QOL and annual income, ageing, pain frequency, and comorbidities ($p < 0.05$).

Table 3 Adolescents' responses for different sub-domains of psychological function

Psychological function	HbSS		HbAS		HbA		
	No	%	No	No	%	No	
Disinterest							
On most days, I do not	27	51.92	128	94.81	185	92.50	< 0.001
On some days, I do	15	28.85	7	5.19	15	7.50	
On most days, I do	10	19.23	0	0.00	0	0.00	
Worry							
On most days, I do not	27	51.92	73	54.07	194	97.00	< 0.001
On some days, I do	11	21.15	45	33.33	6	3.00	
On most days, I do	14	26.92	17	12.59	0	0.00	
Look Perception							
I don't feel I look different	16	30.77	80	59.26	178	89.00	< 0.001
I look different	23	44.23	21	15.56	11	5.50	
I have not though about it	13	25.00	34	25.19	11	5.50	
Feeling sad							
On most days, I do not	27	51.92	90	66.67	197	98.50	< 0.001
On some days, I do	13	25.00	40	29.63	3	1.50	
On most days, I do	12	23.08	5	3.70	0	0.00	
Jealousy							
On most, I do not	39	75.00	101	74.81	182	91.00	< 0.001
On some days, I do	9	17.31	34	25.19	17	8.50	
On most days, I do	4	7.69	0	0.00	1	0.50	

Table 4 Responses of the subjects for different factors of social function

Social function	HbSS		HbAS		HbA		
	No	%	No	%	No	%	
Entertainment							
On most days, not affected	33	63.46	96	71.11	187	93.50	< 0.001
Affected on some days	10	19.23	27	20.00	12	6.00	
Affected on most days	9	17.31	12	8.89	1	0.50	
Teachers support for problems							
Always supportive	39	75.00	115	85.19	179	89.50	< 0.001
Supportive on some days	12	23.08	19	14.07	20	10.00	
On most days not supportive	1	1.92	1	0.74	1	0.50	
Acceptability by friends							
On most days, not affected	43	82.69	119	88.15	195	97.50	< 0.001
Affected on some days	7	13.46	16	11.85	5	2.50	
Affected on most days	2	3.85	0	0.00	0	0.00	
Participation in cultural activities							
Can on most days	25	48.08	112	82.96	194	97.00	< 0.001
Can on some days	9	17.31	21	15.56	5	2.50	
Unable to do on most days	18	34.62	2	1.48	1	0.50	

Discussion

The present study aims to assess the quality of life (QOL) by exploring specific domains most affected in adolescents with SCD and SCT. Several published studies exist on the QOL of individuals affected by SCD (Goldstein-Leever et al. 2020; Bindhani and Nayak 2018; Dale et al. 2011). Most of these studies reported on specific domains reporting average scores of QOL. Furthermore, the focus has mostly been on individuals with SCD, assuming the benign nature of SCT. However, the present study included adolescents with both SCD and SCT to assess their quality of life.

Table 5 Respondent's reaction to different items of health perception (collected only from the individuals with SCD and SCT)

Health perception	SCD		SCT		
	No	%	No	%	
Seriousness of illness					
Like a common cold	17	32.69	85	62.96	< 0.001
More than a common cold	17	32.69	40	29.63	
It is a severe illness	18	34.62	10	7.41	

This study found a significantly lower health-related quality of life (HRQOL) in adolescents with sickle cell genes (HbSS and HbAS) than in healthy adolescents (HbAA), which is similar to the findings of previous studies (Ferro et al. 2021; Hilliard et al. 2020; Dale et al. 2011; Gold et al. 2009; Speechley et al. 2006; Shankar et al. 2005). Individuals with SCT are considered relatively asymptomatic. However, this study revealed that many psychosocial subdomains, for instance, worry about their illness, frequency of angry days, feeling jealousness toward other normal adolescents, and a negative feeling of sadness on some days, were affected almost equally in adolescents having SCT and SCD. This corroborates with a similar study by Patel and Pathan (2005). Also, both SCD and SCT adolescents reported that they were economic burdens on their families because of the extensive hospitalization and health care maintenance, which is another root cause for sadness and depression among the economically poor respondents. This finding is similar to a previous study (Bindhani 2016). The intensity of pain was significantly more severe in SCD than in SCT, which corroborates previous research (Ashorobi et al. 2019; McClish et al. 2005). Further, adolescents with SCT experienced the common cold more frequently (62.96%) compared to their counterparts with SCD, who had a lower incidence rate of about 32.69%.

Table 6 Adolescents' responses for different items of comorbidity

Morbidity	HbSS		HbAS		HbA		
	No	%	No	No	%	No	
Pain frequency							
Never in pain	2	3.85	104	77.04	180	90.00	< 0.001
Sometimes in pain	36	69.23	27	20.00	20	10.00	
Always(most of the time) in pain	14	26.92	4	2.96	0	0.00	
Weakness frequency							
Never feel tired	3	5.77	58	42.96	168	84.00	< 0.001
Sometimes feel tired	32	61.54	68	50.37	29	14.50	
Always feel tired	17	32.69	9	6.67	3	1.50	
Pain intensity reaction							
No sadness	6	11.54	70	51.85	195	97.50	< 0.001
Minimum sadness	8	15.38	40	29.63	3	1.50	
Some sadness	12	23.08	25	18.52	2	1.00	
Cry	13	25.00	0	0.00	0	0.00	
Agony	13	25.00	0	0.00	0	0.00	
Weakness intensity							
No weakness	3	5.77	57	42.22	152	76.00	< 0.001
Minimum weakness	14	26.92	42	31.11	33	16.50	
Some weakness	19	36.54	34	25.19	15	7.50	
Cry	16	30.77	2	1.48	0	0.00	
Agony	0	0.00	0	0.00	0	0.00	

Table 7 Adolescents' responses for different factors of opportunity

Opportunity	HbSS		HbAS		HbA		
	No	%	No	No	%		
Optimism in achieving success (O)							
Optimistic in achieving like peers	11	21.15	75	55.56	160	80.00	< 0.001
Can achieve with difficulty	30	57.69	57	42.22	37	18.50	
Will not be able to achieve	11	21.15	3	2.22	3	1.50	

Table 8 Healthcare cost of families with HbA and without HbS

Healthcare expenditure (% of the family income)	No. of households without HbS($n=482$)		No. of households with HbS ($n=70$)		p-value
	No	%	No	%	
< 10%	351	73.13	28	38.89	< 0.001
10% to < 20%	115	23.96	21	29.17	
20% to < 30%	12	2.50	8	11.11	
30% to < 40%	2	0.42	9	12.50	
> 40%	0	0.00	6	8.33	
Mean healthcare cost (Annually)	**911.4 ± 315.8**		**5182.5 ± 1124**		**< 0.001**

The frequency of blood transfusion received (1.7) was significantly higher in adolescents with SCD than in SCT. However, to the best of our knowledge, no study has reported blood transfusion among sickle cell carriers (SCT). Furthermore, the frequency of hospitalization (2.1) was higher in subjects with SCD than in SCT. Both the frequencies of blood transfusion and hospitalization among SCD-affected individuals were higher in this study compared to the findings of a previous similar study (Patel and Pathan 2005). More importantly, the frequency of blood transfusion and hospitalization was considerably higher among the studied SCT subjects than in the previous study (Patel and Pathan 2005). This study explored the overall QOL score by grouping participants into sickle cell patients, sickle cell carriers, controls with HbS family member(s), and controls without HbS family member(s) for a grassroots-level analysis. Furthermore, the present study reported that breathing problems are common among individuals with SCT, which might have occurred as they generally worked at high altitudes and walked a long distance for schooling. However, no previous study was found related to this type of comorbidities among SCT adolescents.

The percentage of annual household income spent on healthcare overheads ranged from 3.6% to 81.3% in families with sickle cell hemoglobinopathies. In contrast, the expenditure on health in families without sickle cell hemoglobinopathies was 0.8% to 19.2% of their family income. Though previous studies reported a high healthcare expenditure rate in families with sickle cell patients, this was relatively lower compared to the present study (Ngolet et al. 2016; Olatunya et al. 2015). On the other hand, this proportion was more affected in the studied area due to their comparatively lower family income. The substantial difference in healthcare costs among families with sickle cell carriers was observed because families with only sickle cell carriers had lower expenditures compared to families with sickle cell disease. Although the government of Odisha recently implemented a scheme providing 500 rupees for each individual with sickle cell disease, it is negligible compared to their healthcare costs (Bindhani et al. 2021). Some families' healthcare expenditures were out of pocket. In other words, their healthcare care cost exceeded their total monthly income. A similar research described that in most developing countries, the healthcare cost of households with sickle cell hemoglobinopathies is mainly paid through out-of-pocket expenditure (Olatunya et al. 2015). For instance, during the household survey, a family was found where a widow woman had two children; one child had SCD, and the other one had SCT. The mother was also a carrier of HbS with associated comorbidities. Their monthly income was nearly only 1000 Indian rupees, and many times, this amount was very little to treat themselves. Also, many families were found to have the same status in the studied area. Since sickle cell carriers are associated with comorbidities that increase with age, the quality of life of older individuals may be more affected. Therefore, further studies are needed to explore this relationship in more detail.

Conclusion

As expected, the frequency of blood transfusion received (1.7 ± 0.4) was significantly higher in adolescents with SCD than in SCT (0.07 ± 0.3) and healthy controls (0). The notable factor here is that individuals with SCT were found to have undergone blood transfusions in some instances. Furthermore, the frequency of hospitalization (2.1 ± 0.9) was found to be significantly higher among adolescents with SCD. Intriguingly, the frequency of hospitalization among SCT individuals, which is considered a benign state, is significantly higher than that of individuals without HbS. The present study found a significantly lower health-related QOL in adolescents with SCD. However, most psychosocial subdomains, such as worry about illness, frequency of angry days, feeling jealousy towards other normal adolescents, and negative feelings of sadness on some days, are similarly affected in adolescents with SCT and SCD. The overall QOL of SCD individuals is more affected (percentage of affected mean score = 60.93%), followed by SCT individuals (35.63%). Healthy adolescents' QOL is relatively unaffected (13% were affected). Further, healthcare expenditure was significantly higher (3.6% to 81.3% of family income) in

families with HbS than in families without HbS (0.8% to 19.2%) ($p < 0.05$). The overall QOL was affected in both SCD and SCT adolescents. Therefore, equal focus should be given to both SCD and SCT individuals, despite only SCD individuals. The studied population's low socioeconomic and educational status is more like an extra burden on their QOL. Interventions to improve QOL should target the affected domains. Enhancing healthcare facilities and awareness of the disease and its manifestations will help to improve the QOL in adolescents with SCT. Community-based awareness and interventional programs should be implemented to elevate the quality of life in individuals with SCD and SCT.

Author contributions All authors contributed to the study conception and design. Material preparation, data collection, and analysis were performed by BKB. The first draft of the manuscript was written by BKB. JKN reviewed it thoroughly and incorporated his input. All authors read and approved the final manuscript.

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Data availability No datasets were generated or analysed during the current study.

Code availability Not Applicable.

Declarations

Ethics approval The work was approved by the Ethical Committee, Department of Anthropology, Central University of Odisha.

Consent to participate Informed consent was obtained from all patients for being included in the study.

Consent for publication All authors agreed with the content and gave explicit consent to submit.

Competing interests The authors declare no competing interests.

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