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Assessment of Psychosocial Problems in Sickle Cell Phenotypes: An Observational Study from Central India

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ABSTRACT

Background and Objective: Sickle cell disease is a chronic, inherited disease with many complications that worsen the quality of life (QoL) of patients. Although psychosocial problems are common in these patients, they receive the least attention from parents and caregivers. The aim of the present study was to evaluate the psychosocial problems and QoL of children and adolescents with sickle cell disease.

Methods: A cross-sectional study was conducted on 30 patients with sickle cell disease in an Indian tertiary teaching hospital between September 2019 and April 2020. Social and psychological aspects were assessed using the PSC-35 scoring system. To assess the QoL, the current study used a self-designed questionnaire including main psychological, physical, and social domains of the World Health Organization Quality of Life Brief Version (WHOQOL-BREF) scale.

Findings: The mean age of the participants was 8.86 (SD ± 4.54 years) with a range between 2 and 18 years. Among sociodemographic variables, older age, number of hospitalizations, blood transfusions and pain episodes per year were significantly associated with some QoL variables. Although none of the participants in the present study suffered from depression, higher scores on the PSC-35 score were significantly associated with some QoL variables.

Conclusion: Multiple admissions, transfusions, and pain episodes negatively affect the QoL of children and adolescents with sickle cell disease. Because some QoL variables may serve as early predictors of depression, it is recommended to regularly assess the QoL in these children.

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Introduction

Sickle cell disease (SCD) is a chronic haemolytic anemia associated with multisystem morbidity and high mortality [1]. SCD affects more than 250 million people worldwide, with a prevalence of 17% in India [2, 3]. Sickle cell patients and their families are known to have poorer health-related quality of life (QoL) compared to the general population [4- 7]. Many SCD children have psychosocial problems such as low self-esteem, depression, anxiety, and inattention related to frequent hospitalizations, pain crises, community attitudes, and limitations in social, educational, and recreational activities [8]. These psychological complications can occur at any time during these patients' lives and negatively impact their daily living functions and QoL. To provide optimal health care to SCD children, parents and health care providers must recognize the psychosocial issues they face. There is a paucity of research addressing social and mental health aspects of these children. Hence, the aim of the current study was to identify children with sickle cell anemia who had mental health problems using the Pediatric Symptom Checklist (PSC) (HINDI) and to assess the association of different sociodemographic variables and disease severity with QoL. The present study also sought to determine the effectiveness of our simple, self-administered 16-item QoL assessment questionnaire compared with the Pediatric Quality of Life Inventory, version 4.0 Generic Core Scales (PedQL 4.0) [9].

Methods

Design and participants

This cross-sectional study was conducted between September 2019 and April 2020 in Indore, Madhya Pradesh at a tertiary care teaching hospital in central India. Subjects were a convenience sample of 30 children and adolescents aged 2-18 years. Inclusion criteria were all homozygous sickle cell children. Exclusion criteria were children with heterozygous SCD, compound heterozygous SCD, history of psychiatric disorders, developmental delay, mental retardation and autism spectrum disorder.

Data collection

Information on patients was obtained by interviewing parents for sociodemographic details and by reviewing medical records. Social and psychological aspects were assessed using the PSC 35 [10, 11]. Two versions of the 35-item PSC including the parent-completed version (PSC) and youth self-report (Y-PSC) version were used in the ongoing study. The Y-PSC was administered to patients aged ≥11 years. The PSC consists of 35 items classified as "never," "sometimes," or "often" present and scored as 0, 1, or 2, respectively. The total score is calculated by adding the scores for each of the 35 items. For children and adolescents aged 6 to 16 years, a cut-off score of 28 or higher indicates psychological impairment. For children aged 4 and 5 years, the PSC cut-off score is 24 or higher. The cut-off score for the Y-PSC is 30 or higher. Items that are not filled are simply ignored (i.e., the score is 0). If four or more items are left blank, the questionnaire is considered invalid. A positive score on the PSC or Y-PSC suggests depressive symptoms. To assess the QoL among SCD patients, we developed a self-designed 16-item questionnaire including the main psychological, physical, and social domains of the WHOQOL-BREF scale [12].

Statistical analysis

Statistical analysis was performed using SPSS. The association between sociodemographic details and QoL variables was calculated by applying the chi-square test. We also compared QoL and its subscales with the depression scale (PSC -35 score).

Results

Of the 30 SCD patients who participated in the study, 50% were male. The mean age of the participants was 8.86 ± 4.54 years with a range of 2-18 years. The socio-demographic characteristics of the patients are represented in Table 1. The majority of participants (76.6%) belonged to the middle class. The frequency of ≥3 hospitalizations and

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blood transfusions per year was 16.7% (5 cases) and 13.3% (4 cases), respectively. In the present study, ≥3 episodes of pain per year were observed in only 3.3 % of the study participants. In the PSC scoring system, we had the highest score of 9 in 3 cases (10%), representing no major psychological problems in the study group. Most of the children (63.3%) scored only 1-3. In our study, none of the participants had enough points to reach the cut-off point for psychosocial problems in the PSC score. The frequency of various responses to the QOL variables among the study participants is illustrated in Table 2.

We compared some sociodemographic variables with QoL variables. Among QoL variables, feelings of isolation (p=0.02), limitation of physical activity (p=0.04), and sleep disturbance (p=0.04) were significantly associated with older age. Similarly, the number of hospitalizations per year among study participants was significantly associated with the variables of isolated feelings (p=0.02), severe activity restriction (p=0.02), sleep disturbance (p=0.01), and loss of appetite (p=0.01). Blood transfusions and pain episodes per year were statistically significantly associated with all QoL variables (p<0.05). In contrast, the association of gender, socioeconomic status, outpatient department (OPD) visits, and short stature with QoL variables was not significant. Higher scores on the PSC score were significantly associated with some QoL variables. Children who experienced physical and psychosocial problems, such as school absenteeism, limitation of physical activity, sleep disturbances, pain in the past 4 weeks, and negative feelings, had higher PSC scale scores and were statistically significant (p<0.05), whereas other variables were not significant (Table 3).

Table 1. Sociodemographic and clinical characteristics (N=30) of the study participants

Variables	Amount		
Ages in years (mean ±SD)	≤5 (n=9)	5-10 (n=8)	>10 (n=13)
	3.6±1.05	7.3±1.3	14±1.1
Caste: N (%)	ADIWASI	05 (16.7)	
	BALAI	04 (13.3)	
	BALNI	02 (6.7)	
	BHEEL	05 (16.7)	
	BHILLALE	01 (3.3)	
	GOND	03 (10)	
	others	10 (33.3)	
Region: N (%)	Urban	15 (50)	
Socioeconomic status*: N (%)	Upper lower	07 (23.3)	
	Lower middle	13 (43.3)	
	Upper middle	10 (33.3)	
No. of times hospitalized per year: N (%)	Nil	<3	≥3
	1(3.3)	24 (80)	5 (16.7)
No. of transfusions received per year: N (%)	Nil	<3	≥3
	0	26 (86.6)	4 (13.3)
No. of OPD visits per year N (%)	Nil	<3	≥3
	0	26 (86.6)	4 (13.3)
No. of severe pain episodes per year N (%)	Nil	<3	≥3
	4 (13.3)	25 (83.3)	1 (3.3)
Height for age (percentile-NCHS) for age 5-17 years N (%)	<3rd	01 (4.76)	
	3rd – 10th	29 (95.2)	
BMI for ages 5-17 years N=21(%)	>50th	03 (14.3)	
	25th -50th	03 (14.3)	
	10th -25th	05 (23.8)	
	5th -10th	05 (23.8)	
	3rd -5th	02 (9.5)	
	<3rd	03 (14.3)	
Nutritional status for under 5 years of age N=9(%)	MAMα	09 (100)	
	SAMβ	0	

*On basis of the Modified Kuppuswami scale,
α Moderate acute malnutrition,
β severe acute malnutrition

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Table 2. Responses to Quality of Life (QoL) variables

N	Psychosocial Domain			N	Physical Domain		
	Item	N=30	%		Item	N=30	%
1	**Experienced negative treatment**			1	**Vigorous activities**		
	Yes	0	0		Yes, a Lot	0	0
	No	30	30		Yes, a Little	4	13.30
					No	26	86.70
2	**Experienced any negative attitudes**			2	**Household works**		
	Yes	0	0		Yes, a Lot	0	0%
	No	30	30		Yes, a Little	0	0.00%
					No	30	100.00%
3	**Isolated feeling in a child**			3	**Climbing flights of stairs**		
	Yes, sometimes	3	10		Yes, a Lot	0	0
	Yes, a lot	0	0		Yes, a Little	0	0.00
	No	27	90		No	30	100.00
4	**Awareness in society**			4	**Routine activities**		
	Yes	30	100		Yes, a Lot	0	0
	No	0	0		Yes, a Little	0	0.00
					No	30	100.00
5	**Awareness in school**			5	**Sleep disturbance**		
	Yes	30	100		Yes, a Lot	0	0
	No	0	0		Yes, a Little	4	13.30
					No	26	86.70
6	**Social program participation**			6	**Loss of appetite**		
	Yes	30	100		Yes, a Lot	0	0
	No	0	0		Yes, a Little	2	6.60
					No	28	93.40
7	**Negative feelings in a child**			7	**Pain episodes during the past 4 weeks**		
	Yes, a lot	0	0		None	22	73
	Yes, sometimes	8	36		Mild	6	20
	No	22	64		Moderate	2	6.6
8	**Negative feelings in parents**			8	**School absenteeism**		
	Yes	0	0		Yes, a Lot	3	10
	No	30	100		Yes, a Little	4	13.30
					No	23	76.60

Table 3. Association between 35-item PSC and QoL variables

Variable	Response	PSC-35 SCORE			Total N=30	P value
		1-3	4-6	7-9		
Negative Feelings	No	16	5	1	22	0.00*
	Sometime	0	3	5	8	
Isolated Feelings	No	19	6	2	27	0.05
	Sometime	0	2	1	3	
Appetite Loss	No	19	7	2	28	0.07
	Sometime	0	1	1	2	
School Absenteeism	No	16	6	1	23	0.01*
	Sometime	3	1	0	4	
	Yes	0	0	1	3	
Vigorous Activity Limitation	No	19	6	1	26	0.00*
	Sometime	0	2	2	4	
Sleep Disturbance	No	19	6	1	26	0.00*
	Sometime	0	2	2	4	
Pain in last 4 Week	No Pain	16	6	0	22	0.02*
	Mild	3	1	2	6	
	Moderate	0	1	1	2	

*p value<0.05 statistically significant

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Discussion

In the present study, out of the total 30 children, 50% of the study participants were males, whereas Kambasu et al. [13] and Habeeb et al. [14] found 40% and 62.3% males in their study, respectively. The mean age in the present study was 8.86 ± 4.54 years with an age range of 2-18 years, whereas the mean age of SCD children in the case-control study by Sehlo and Kamfar [15] was 11.93 ±1.72 years with an age range of 10-15 years. Many studies conducted worldwide with SCD patients included people of different ages (Table 4).

In the present study, it was observed that no participant had depression and psychosocial symptoms that reached to a treatable limit, similar to Noll et al., [18] while Sehlo MG [15] observed depression in 13% of the 60 study participants. This could be due to differences in sample size, disease severity, or different measurement tools, as they used the 27-item Children's Depression Inventory (CDI) scale to assess depression. Another reason for the absence of depressive symptoms in our study participants could be good social support from their teachers, parents, and friends, as suggested by QOL variables. Such contradictory results require further studies on this aspect with a larger sample size.

Some QoL variables such as feelings of isolation (p=0.02), physical activity limitation (p=0.04), and sleep disturbance (p=0.04) were significantly associated with older age, similar to the findings of Kambasu et al. [13] who observed that children aged greater than 12 years had statistically significant lower scores in the emotional and psychosocial domains of the health-related QoL (HRQoL) than children aged less than 12 years. Adeboyega et al. [19] also found that older age was associated with lower HRQoL. In contrast, Sehlo and Kamfar [15] found no age as a predictor of low HRQoL. Previous studies found significant differences in QoL related to gender [4, 13, 20]. In the present study, consistent with other studies, gender had no effect on patients' QoL [5, 15, 21, 22]. In contrast to Panepinto JA et al., [23] no statistically significant association between SES and QoL was found in the present study. Statistical analysis revealed that the number of blood transfusions and pain episodes per year was significantly associated with several domains of QoL, similar to previous studies [5, 13-15, 17]. These clinical conditions tend to worsen the QoL of SCD patients.

It is well known that the presence of social support serves as a buffer for children at risk. Our study participants had good social support from their parents, friends, and school as suggested by their responses in the psychosocial domains, whereas Kambasu et al. [13] reported that SCD negatively affected school functioning, physical functioning and emotional QoL. Thus, it is concluded that the results of our simple and less time-consuming 16–item scale are comparable to other HRQoL scales. In the present study, although no participant was found to have depression, higher scores on the PSC scale were significantly related to negative feelings, school absenteeism, limitation of physical activity, sleep disturbances, and pain in the past four weeks. This means that all of these factors may be early predictors of the development of depression in children and adolescents with SCD. Early counselling and treatment of these variables may mitigate the development of depression and its effect on QoL.

Table 4. Study population in various studies

Author and year of publication	Sample size	Age range (in Years)
Kambasu et al, 2019 [13]	140	8 -17
Habeeb et al 2015 [14]	130	6 -18
Sehlo and Kamfar, 2015 [15]	60	10 -15
Ogre Sc et al ,2018 [16]	309	11-19
Al Jaouni et al, 2013 [17]	115	2- 48
Present study	30	2-18

Limitations of the study

The limitations of this study were a hospital-based cross-sectional study with a smaller sample size; therefore, a community–based, longitudinal study with a larger sample size is needed to address the psychosocial and physical problems of children and adolescents with SCD. Community-based studies of SCD can also address the needs of those children and adolescents who receive irregular medical care for a variety of reasons.

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Conclusion

For comprehensive management of SCD patients, both physical and mental health should be adequately addressed. However, parents and medical professionals pay more attention to physical health and neglect psychosocial aspects. The results of the present study showed that frequent blood transfusions, more episodes of pain, more hospitalizations, and increasing age negatively affected the QoL of children and adolescents with SCD. The QoL assessment is recommended for each SCD patient to improve their mental health, because some QoL variables may be early predictors of depression.

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Ethical approval

The Nuremberg Code of Ethics was applied to obtain institutional ethics committee approval (Ethical code-SAIMS/RC/EC/2019/12 Dated 2/12/2019) Written informed consent was obtained from all study participants and their parents.

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Conflict of interest

There are no conflicts of interest.

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