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Quality of life among individuals with sickle cell disease: A study from Koraput district, Odisha

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Abstract
Introduction: Sickle cell disease (SCD) is a genetic disorder with an estimated 5,200 live births each year is a major public health problem in India. Although SCD has been described in India in numerous ethnic groups, it is most prevalent in tribal community. Prevalence of Sickle Cell gene is 5 to 34 % in tribal community, who have a high prevalence of socio-economic disadvantage and are frequently medically underserved.
Objectives: The present study was conducted with the aim of assessing QOL and to explore specific domains or factors that are most affected in individuals with sickle cell disease with respect to normal people.
Methods: A qualitative case study approach has been employed among 43 SCD affected individuals in age group of 4-48 in 9 villages of Koraput district which is one of the most backward districts of the state and predominantly tribal populated one. The villages were selected accordingly the availability of SCD patients in that village. More priority was given to those villages which have more numbers of SCD patients and more remote, where medical facilities are not easily assessable. Among the selected sample (n=43), children (n=15), where all of them were students, were separated for a categorized study. The quality of life (QOL) was assessed using multidimensional interviews and schedules. Subsequently, Scoring Scales were developed and modified for an intensive study.
Results: Result reveals that all factors like physical, psychosocial and cognitive were affected. Their everyday activities like working or schooling, vocational achievement perception, entertainment and participation in cultural activities, and socio economic factors were also affected. Most of the time they feel sad, scared and disinterest. The intensity of weakness and pain was greater in SCD patients as compare to the normal children.
Conclusion: Over all QOL is affected in individuals with sickle cell disease (SCD). Interventions to improve QOL should target the affected items. Improving awareness of the disease and its manifestation will help to alleviate the psychosocial affliction of individuals with SCD.

Keywords: sickle cell disease (SCD), quality of life (QOL), health activities, students, Koraput

Introduction
SCD is one of the most common monogenic disorders globally with an autosomal recessive inheritance. Sickle cell disease (SCD is a major public health problem with an estimated 5,200 live births each year in India.
James Herrick (1910) [1] in his work, "Peculiar elongated and sickle-shaped red blood corpuscles in a case of severe anaemia", was confirmed the elongated shape of a red blood cell. Herrick is the first person to discover SCD. Adeyemo *et al*. (2015) [2] conducted a cross sectional study on "health related quality of life and perception of stigmatization in adolescents living with sickle cell disease in Nigeria". The study was conducted on 160 adolescents, which included 80 participants with SCD and 80 participants without SCD. Health surveys were distributed to participants to measure how stigmatization affects the HRQL. The results of the study shows that participants with SCD have lower HRQL than those without the SCD.
In this aspect of the literature review, previous researchers have done very little research and less research has been conducted in the last five years regarding quality of life with SCD. There is no study conducted in Koraput district of Odisha, where the prevalence of SCD is more.
The present study was conducted with the aim of assessing QOL and to explore specific domains or factors that are most affected in individuals with sickle cell disease with respect to normal people in Koraput district of Odisha.

Materials and Methods
Koraput was purposefully selected for study because this district is one of the most backward districts of the state and predominantly tribal populated one and there is not a single work done before regarding sickle cell anaemia.
The villages (n=9) were selected accordingly the availability of SCD patients in that village. More priority was given to those villages which have more numbers of SCD patients and more remote, where medical facilities are not easily assessable.
A qualitative case study approach has been employed among 43 SCD affected individuals in age group of 4-48 in Koraput district of Odisha. Among the selected sample (n=43), children (n=15), where all of them were students, were separated for a categorized study. The quality of life (QOL) was assessed using multidimensional interviews and

schedules. Subsequently, Scoring Scales were developed and modified for an intensive study.

A focus group discussion with parents and children with SCD was conducted to identify domains for the QOL scale. In addition, preliminary analyses were conducted to examine the internal consistency of the study measures (e.g., PPQ, PedsQL, SCD-QoL, PCQ, and MIBI) with the obtained sample. For data analysis purpose SPSS and MS Excel software tools were used.

Results and Discussion

Families Experiencing Sickle Cell Anaemia

A total of 43 patients diagnosed with sickle cell anaemia were found in the sample of 28 families.

Table 1: Total number of SCD affected with sex distribution

SCD affected individuals	Number	Percentage
Male	19	44.19
Female	24	55.81
Total(n)	43	100.00

Table-1 shows the frequency of male and females affected with SCD. Out of 43 patients 19 (40.2%) are male and 24 (55.8%) are female.

Table 2: Age-Sex distribution of SCD affected individuals

Age group (in year)	Male		Female		Total	
	Number	Percentage	Number	Percentage	Number	Percentage
0-5	2	10.53	0	0	2	4.65
6-10	2	10.53	7	29.17	9	20.93
11-15	3	15.79	2	8.33	5	11.63
16-20	2	10.53	5	20.83	7	16.28
21-25	1	5.26	4	16.67	5	11.63
26-30	3	15.79	1	4.17	4	9.30
31-35	3	15.79	2	8.33	5	11.63
36-40	1	5.26	2	8.33	3	6.98
41-45	1	5.26	1	4.17	2	4.65
46-50	1	5.26	0	0	1	2.33
Total	19	100	24	100	43	100

In table-2, the individuals affected with SCD were categorized in 10 age groups of class interval 5. The descriptive statistics of age shows that the minimum age of the sample is 3 years and the maximum aged individual is 48. (Mean=21.51, Standard deviation=12.23). And maximum numbers of patients are falls under the age group 6 to 10 years, i.e. 21% of the total SCD patients.

Table 3: Caste- Sex distribution

Caste	Male		Female		Total	
	Frequency	%	Frequency	%	Frequency	%
SC	14	73.68	22	91.67	36	83.72
ST	5	26.32	2	8.33	7	16.28
Total	19	100	24	100	43	100

Table-3 explores that in my sample data more numbers of SCD affected individuals belongs to SC category, which constitutes 83.72% of the total patients. And this is followed by the ST category i.e. 16.28%. SC females are more than the ST males by 17.99%. But in case of ST category, males are more than female with a difference of 17.99.

Table 4: Gender wise Distribution of Religion SCD patients

Religion	Male		Female		Total	
	No.	%	No.	%	No.	%
Hindu	12	63.16	20	83.33	32	74.42
Christian	7	36.84	4	16.67	11	25.58
Total	19	100	24	100	43	100.00

Similarly, there are more numbers of Hindu patients the sample. Out of 43 patients 32 (74.42%) individuals are Hindu and 11 (25.58%) individuals are of Christians. (See table-4)

Table 5: Educational Level of the Patients

Educational Level		Male		Female		Total	
		No.	%	No.	%	No.	%
School level	up to 5^{th}	5	26.32	10	41.67	15	34.88
	6^{th} -10^{th}	9	47.37	13	54.17	22	51.16
Intermediate		2	10.53	0	0.00	2	4.65
Others		1	5.35	0	0.00	1	2.33
Illiterate		2	10.53	1	4.17	3	6.98
Total		19	100	24	100	42	100

The samples are taken basically from the remote areas of the Koraput district. Most of the people are illiterate or under primary level. Table-5 shows the educational level of the patients. A large number of SCD affected individuals are under 10^{th} standard. Out of 43, 37 (86.04%) individuals are under 10^{th} standard. The intermediates and illiterates constitute 4.65% and 6.98% respectively. There is nobody has a graduate degree in the sample.

Table 6: Occupational distribution of patients

Occupation	Male		Female		Total	
	Frequency	%	Frequency	%	Frequency	%
Housewife	0	0.00	11	45.83	11	25.58
Labour	5	26.32	1	4.17	6	13.95
Farmer	2	10.53	0	0.00	2	4.65
Student	7	36.84	8	33.33	15	34.88
Employed (Private)	2	10.53	0	0.00	2	4.65
Others	3	15.79	4	16.67	7	16.28
Total	19	100.00	24	100.00	43	100.00

By analysing the above table (table-6), indicates that most of them are unemployed because many individuals are students. Out of 43 affected individuals 19 individuals, constituting 44.2% are unemployed. That implies those unemployed are totally depending upon their families for their treatment, which ultimately decreases the family's economic condition.

Table 7: Household Income per annum

Annual household income (in rupees) = *x*	Number of household	Percentage
x ≤ 50,000	9	20.93
50,000<x<1,00,000	33	76.74
x≥1,00,000	1	2.33

Average household income $\bar{x}$ = 66,907 /- (SD=13636.11)

Table-7 explores the house household income of different families. All the families have a very low annual income. I found that the annual income ranges from lower limit 40,000 rupees to upper limit 1, 00,000 rupees only. The average household income is 66,907 rupees only *(SD=13636.11)*, which is a very low amount to survive in the present world for a normal family. But here extra burden is to spend a larger part of money for the medication of the patient (s) in the family.

Insurance: Out of the twenty-eight families in the sample, only six (13.95%) patients in four (14.3%) families have insurance coverage. One of the families indicated an increase in the rate of insurance because of the patient's illness. Those families w1th medical insurance coverage however, explained that the insurance was obtained through employment benefits. Life 1nsurance for the patients was presented as a problem for these families, in that none of the families were able to obtain life insurance for the patient with sickle cell anaemia.

Paediatric Quality of Life Inventory

Table 8: Paediatric Quality of Life Inventory

About my Health and Activities (problems with....)	Never		Almost never		Some-times		Often		Almost Always		No Ans.	
	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
It is hard for me to walk more than one block.	1	2.33	12	27.91	30	69.76	0	0.00	0	0.00	0	0.00
It is hard for me to run.	1	2.33	11	25.58	28	65.12	3	6.98	0	0.00	0	0.00
It is hard for me to do sports activity or exercise.	0	0.00	7	16.28	31	72.09	2	4.65	2	4.65	1	2.33
It is hard for me to lift something heavy.	0	0.00	1	2.33	38	88.37	3	6.98	0	0.00	1	2.33
It is hard for me to take a shower or bath by myself.	32	74.42	0	0.00	10	23.26	0	0.00	0	0.00	1	2.33
It is hard for me to do chores around the house.	1	2.33	11	25.58	16	37.21	14	32.56	0	0.00	1	2.33
I have low energy.	0	0.00	8	18.60	30	69.77	5	11.63	0	0.00	0	0.00

Table 9: Paediatric Quality of Life Inventory

About My Feelings (problems with....)	Never		Almost never		Some-times		Often		Almost Always		No Ans.	
	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
I feel afraid or scared.	7	16.28	22	51.16	10	23.26	3	6.98	0	0.00	1	2.33
I feel sad or blue.	7	16.28	22	51.16	10	23.26	3	6.98	0	0.00	1	2.33
I feel angry.	0	0.00	12	27.91	30	69.77	0	0.00	0	0.00	1	2.33
I have trouble sleeping.	0	0.00	22	51.16	20	46.51	0	0.00	0	0.00	1	2.33
I worry about what will happen to me.	0	0.00	0	0.00	37	86.05	5	11.63	0	0.00	1	2.33

Table-8 and table-9 shows a list of things that might be a problem for SCD affected individuals regarding to theirs health activities and feelings respectively. And each individual in the sample asked to tell how much of a problem each one has been during the past one month.

Paediatric Quality of Life Inventory for students

Table 10: Paediatric Quality of Life Inventory for students

How I Get Along With Others (problems with....)	Never		Almost never		Some-times		Often		Almost Always		No Ans.	
	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
I have trouble getting along with other kids.	8	53.33	7	46.67	0	0.00	0	0	0	0	0	0
Other kids do not want to be my friend.	13	86.67	2	13.33	0	0.00	0	0	0	0	0	0
Other kids tease me.	15	100.0	0	0.00	0	0.00	0	0	0	0	0	0
I cannot do things that other kids my age can do.	3	20.00	6	40.00	6	40.00	0	0	0	0	0	0
It is hard to keep up when I play with other kids.	3	20.00	6	40.00	6	40.00	0	0	0	0	0	0

Table 11: Paediatric Quality of Life Inventory for students

About School (problems with....)	Never		Almost never		Some-times		Often		Almost Always		No Ans.	
	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
It is hard to pay attention in class.	2	13.33	8	53.33	5	33.33	0	0	0	0	0	0
I forget things.	10	66.67	4	26.67	1	6.67	0	0	0	0	0	0
I have trouble keeping up with my school work.	6	40.00	4	26.67	5	33.33	0	0	0	0	0	0
I miss school because of not feeling well.	0	0.00	0	0.00	15	100.0	0	0	0	0	0	0
I miss school to go to the doctor or hospital.	0	0.00	0	0.00	15	100.0	0	0	0	0	0	0

Table-10 and table-11 describes about problems of students relating to their schools and friends. I found 15 numbers of students in my sample, where 7 (46.67%) students are males and 8 (53.33%) students are females. In every negative question like, "Does other kids tease you?", "Don't other kids want to be your friend?", when I asked the students those affected with SCD, I found that, all most all answers were no.

Table 12: Paediatric Quality of Life Inventory rating (for students)

Problems with.... (n=15)	Mean Rating	Std. Deviation
I have trouble getting along with other kids	0.47	0.52
Other kids do not want to be my friend	0.27	0.46
Other kids tease me	0.13	0.35
I cannot do things that other kids my age can do	1.13	0.83
It is hard to keep up when I play with other kids	1.20	0.77
It is hard to pay attention in class	1.33	0.72
I forget things	0.47	0.74
I have trouble keeping up with my schoolwork	1.20	0.86
I miss school because of not feeling well	2.00	0.00
I miss school to go to the doctor or hospital	2.00	0.00

[Paediatric Quality of Life Inventory Rating Scale:- Never (0) Almost never (1) Sometimes (2) Often (3) Almost Always (4)]

Table-12 represents the paediatric quality of life inventory ratings. 15 students were included for this study and every one was asked 10 questions. For each question an average rating was calculated for 15 students, which can be called as the intensity of the paediatric quality of life of the students. To get the rating a 5-scale rating scale was developed which was given below.

Some clinical problems like joint pain, chest pain, stomach pain, back pain, body ache, fever, irritability, general weakness, fatigue & giddiness, pallor, blood transfusion reported among the sickle cell patients in common.

Results shows that all factors like physical, psychosocial and cognitive were affected among SCD affected individuals. Their everyday activities like working or schooling, vocational achievement perception, entertainment and participation in cultural activities, and socio economic factors were also affected. Most of the time they feel sad, scared and disinterest. The intensity of weakness and pain was greater in SCD patients as compare to the normal children.

Conclusion and Suggestions

The purpose of this study was to assess QOL and to explore specific domains or factors that are most affected in individuals with sickle cell disease with respect to normal people in Koraput district of Odisha.

In conclusion, over all QOL is affected in individuals with sickle cell disease (SCD). Interventions to improve QOL should target the affected items. Improving awareness of the disease and its manifestation will help to alleviate the psychosocial affliction of individuals with SCD.

In response to collected data from the families experiencing sickle cell anaemia, and from the search of literature', the following recommendations are made:

- In the studied area it was found that all most all persons marry without testing their blood and also they prefer to marry their nearer people and relatives (consanguineal marriage) that lead to a greater probability to be a carrier or affected with SCD. Hence village wise awareness programmes should be implemented in the sickle cell belts.
- All persons experiencing genetic disease have the option to choose genetic counselling and sufficient community education to know they have this choice.
- That all genetic counselling include supportive family therapy and education over a sufficient period of time, so the family can integrate the information into their own life systems.
- That community educative effort is initiated to help parents learn and- understand more about sickle cell anaemia.

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