ORIGINAL RESEARCH ARTICLE
pISSN 0976 3325 | eISSN 2229 6816
Open Access Article
www.njcmindia.org
DOI: 10.5455/njcm.20190415043523

Awareness Regarding Key Aspects of the Sickle Cell Disease and Trait among the Affected In a Tertiary Care Hospital in South Gujarat

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Financial Support: None declared
Conflict of Interest: None declared

How to cite this article:
Nagar SS, Patel HB. Awareness Regarding Key Aspects of the Sickle Cell Disease and Trait among the Affected In a Tertiary Care Hospital in South Gujarat. Natl J Community Med 2020;11(3):112-117

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Date of Submission: 15-04-19
Date of Acceptance: 23-03-20
Date of Publication: 31-03-20

ABSTRACT

Introduction: Sickle cell disorder is an autosomal recessive condition, in which inheritance of an affected gene from both parents results in a disorder. SCD is more prevalent among the socio- economically disadvantaged and medically underserved communities mainly in Tapi District of Gujarat.

Objectives: 1.To study Awareness Regarding Sickle Cell Disease or Trait among the affected 2.To find out the association between the awareness level and the socio-demographic profile of the subjects.

Methodology: Descriptive Cross-sectional study was carried out in Janak Smarak hospital in Vyara Town, Tapi District, for a period of one year and minimum of 75 persons with SCD and 150 persons with SCT were taken for the study.

Results: The overall Knowledge regarding disease was averaging only at 15 %. The categorization of subject having "Good" or "Not so good" among the subject were predominantly poor with a majority of them showing Not so Good condition (84%) whereas only 16% showed good Knowledge.

Conclusion: The overall Knowledge regarding disease was averaging only at 15 % which showed that the awareness seems to be grossly deficit even among the affected. It hence shows that the counseling of the disease among the affected was not adequate. So Counselling & IEC can play a very important role to eliminate the disease from the community.

Key words: Sickle cell Disease, Sickle cell trait, Tribal Community, Awareness.

INTRODUCTION

Sickle cell disease (SCD) is genetic blood disorder that affects the Haemoglobin in RBC. Sickle cell disorder is an autosomal recessive condition, in which inheritance of an affected gene from both parents results in a disorder while inheritance of one abnormal gene results in a healthy carrier.[1]It is characterized by Vaso-occlusive pain crises, risk for pneumococcal infections, acute chest syndrome, and stroke and organ failure and is associated with substantial morbidity and premature mortality.[2]The main reasons for mortality among sickle cell disease patients are infection, acute splenic sequestration, severe anaemia and haemolytic crisis.[1]

According to hospital based epidemiological survey, the prevalence of sickle cell gene is observed to be 0-18 percent in northern- eastern India, 0 to 33 percent in western India,22.5 to 44.4 percent in central part of India and 1 to 40 percent in southern India and the gene frequency of Hb-S varies between 0.031- 0.41. [3]Another survey done by the In-

dian Red Cross Society in Gujarat, where nearly 1, 68,498 tribal individuals from 22 districts were screened, the overall prevalence of sickle cell carriers was found to be 11.37 per cent. [4, 5] Some studies have also documented high prevalence of sickle gene in various tribal communities of Gujarat that include Bhils and Dhodias of Panchmahal, Dublas, Naikas, Koli, Dhanka, Gamit, Vasava, Bariya, Varli, Vaghari, Kukna, Halpati, & Chaudhari. Tribal population accounts for 15 % of the total population of Gujarat and are distributed in various districts of the state such as Sabarkantha, Banaskantha, Panchmahal, Vadodara, Narmada, Bharuch, Surat, Valsad, Dang and the surrounding UTs like Div-Daman. [6] In Tapi District of Gujarat, Sickle Cell Disease is more prevalent among the socio- economically disadvantaged and medically underserved communities.

During 2005-06, Govt. of Gujarat has taken concrete steps for establishing a Comprehensive Sickle Cell Program in four districts of south Gujarat, viz.; Surat, Navsari, Valsad and Dang which has now extended to all 12 tribal districts of Gujarat in 2008. This program is based on public-private partnership. Gujarat Sickle Cell Anaemia Control Society has been formed under society registration act 21 of 1860 to integrate all the activities done by different departments and NGOs in Gujarat for prevention and control of Sickle Cell Anaemia under one umbrella. [7]

So it necessary to educate the people regarding sickle cell disease and sickle cell trait among people so we can prevent it.

OBJECTIVES OF THE STUDY

The research was undertaken to study Awareness Regarding Sickle Cell Disease or Trait among the affected and to find out the association (if any) between the awareness level and the socio-demographic profile of the subjects

MATERIALS & METHODS

The study was carried out in Janak Smark hospital in Vyara Town, Tapi District, Gujarat. It was a Descriptive Cross-sectional study from March 2014 to February 2015 i.e. for a period of one year. After getting permission from the institutional ethics committee of Sumandeep University, Piparia, individuals having sickle cell disease or sickle cell trait (diagnosed at Janak Smarak Hospital, Vyara) were contacted to explain the purpose of the study and nature of his / her participation. Data regarding their demographic and social profile and their knowledge levels was collected after their written consent using a pre-designed and pre-tested questionnaire by directly interviewing them. Only those who consented for the study were included as subjects to collect the data further. Prior permission from the hospital authorities was also sought to carry out the study.

All the 225 patients, who diagnosed with Sickle cell Disease and Sickle cell trait between the period of March 2014 to February 2015, were included as part of the study. This was hence taken as the sample size for the study.

Categorization of subject as having "Good" or "Not So good" knowledge for various aspects of SCD & SCT

Variables	Score
Disease Transmission (Hereditary Cause)	2
Sign & Symptoms	2
Current Haemoglobin Status	2
Sickle Crisis Identification	3
Blood Transfusion in case of emergency	3
Nearest blood bank with respect to their residential location in case of Emergency	3
Good	>11
Not so good	<11

To assess the, knowledge regarding disease, six variables were taken into consideration. Out of the total 6 variables, 3 were considered to be of primary knowledge and the remaining 3 of core knowledge regarding sickle cell anaemia hence, the subjects with positive responses in the former category were given a score of **2** and a score of **3** was accorded to positive responses in the latter 3 variables. For negative responses, score of **1** was given for all the variables. To categorize the subjects into having **"Good"** and "**Not so good"** knowledge, the total scores for each study subjects was calculated. Those scoring more than the score of 11 (Average of maximum and minimum scores of 15 and 6) were categorized as having "Good" knowledge and those scoring below were categorized as having "Not so good" knowledge" levels.

The data collected was analyzed using MS Excel 2007 and suitable statistical tests were applied at 5% level of significance.

RESULTS

All the 225 patients, who were diagnosed with Sickle cell Disease and Sickle cell trait, had consented to be part of the study and as they were very eager to know about Sickle cell Disease and Sickle cell trait.

Table 1 suggests that proportion of females affected were more than that of their male counter-

Table 1: Socio demographic profile of the subjects

Variable	Female (n=132)	Male (n=93)	Total (n=225)
Age (Years)			
1 – 5	4 (3.03)	2 (2.15)	6 (2.66)
17-Jun	33 (25)	34 (36.55)	67 (29.78)
18-45	81 (61.37)	48 (51.61)	129 (57.34)
46- 100	14 (10.6)	9 (9.67)	23 (10.22)
Caste			
Gamit	99 (75)	66 (70.96)	156 (69.33)
Chaudhari	39 (29.54)	23 (24.73)	62 (27.55)
Others*	3 (2.27)	4 (4.3)	7 (3.11)
Marital Status			
Married	72 (54.54)	34 (36.55)	106 (47.74)
Unmarried	56 (42.41)	59 (63.43)	115 (51.38)
Widow/widower	4 (3.03)	0 (0)	4 (1.77)
Religion			
Hindu	123 (93.18)	86 (92.47)	209 (92.88)
Christian	9 (6.81)	7 (7.52)	16 (7.11)
Education			
Illiterate	25 (18.93)	7 (7.52)	32 (14.22)
Primary	34 (25.75)	35 (37.63)	69 (30.65)
Secondary	40 (30.3)	23 (24.73)	63 (28)
Higher Secondary	18 (13.63)	10 (10.57)	28 (12.44)
Diploma/ITI/PTC	1 (0.75)	1 (1.07)	2 (0.88)
Graduate	12 (9.09)	15 (16.12)	27 (12)
PG/Diploma	2 (1.51)	2 (2.15)	4 (1.77)
Occupation			
Student	55 (41.66)	52 (55.91)	105 (46.65)
Housewife	67 (50.75)	0 (0)	67 (29.77)
Farmer	6 (4.54)	24 (25.8)	30 (13.33)
Job	0 (0)	5 (5.37)	5 (2.22)
Teacher	5 (3.78)	5 (5.37)	10 (4.44)
ANM	1 (0.75)	0 (0)	1 (0.44)
Shopkeeper	1 (0.75)	0 (0)	1 (0.44)
Driver	0 (0)	1 (1.07)	1 (0.44)
Social Class* calculated as per capita family income**			
CLASS -I	42 (31.8)	31 (33.3)	73 (32.4)
CLASS - II	77 (58.3)	50 (53.8)	127 (56.4)
CLASS - III	12 (9.1)	9 (9.7)	21 (9.3)
CLASS - IV	0 (0)	2 (2.2)	2 (0.9)
CLASS - V	1 (0.8)	1 (1.1)	2 (0.9)

* "Others" include Valvi and Kokani castes.
** Social class as per Modified B.G. Prasad`s Classification of 2015 (AICPI= Rs. 816/-) [17]

Table 2: Knowledge regarding various aspects of SCD and SCT

Knowledge Aspect	Aware	Not aware
Disease Transmitted (Hereditary Cause)	11	89
Sign & Symptom	24	76
Current Hemoglobin status	6	94
Sickle Crisis Identification	18	82
Blood Trannsfusion in case of emergency	16	84
Nearest blood bank with respect to their residential location in case of emergency	15	85

parts. Majority of the participants belonged to Gamit caste and most of them were Hindus. Majority of the participants were married. Majority of subjects among both males and females were educated up to secondary with more proportion of illiterate among females was more than males. Most of the patients were from Social Class 1 and 2.

Table 2 showed that among the study subjects, it was observed that only 11 % of the subjects were aware about hereditary route of disease transmission, and about 24 % of the subjects had knowledge regarding the various signs and symptoms of the disease. Only 6 % of the subjects knew their current hemoglobin status and only18% of the subjects knew how to identify an episode of sickle cell crisis. When asked about the availability of blood transfusion services to the SCD and SCT individuals, only 16 % of the subject knew regarding the requirement of blood transfusion in case of emergencies like sickle cell crisis. For awareness regarding nearest blood bank with respect to their residence only 15 % of the subject knew about it.

It was noted from **Table 3** that in the study, most (63=84%) of Sickle Cell Diseases patients and (141=94%) of the Sickle Cell Trait subjects knew about the program run by the Government. The knowledge of program among sickle cell trait patients was significantly higher as compared to the sickle cell disease patients.

It was seen from **Table 4** that, to find out the impact of Socio Demographic Profile on the knowledge, all of the factors were analysed & association was sought using chi squared test at 5% level of significance. It was seen that 60% of the patients in the age group between 18-45 years Age (X^2 *value= 12.18; p<0.01);* 67.56% of the Gamit Caste *(X^2 value= 9.024; p<0.05*); 70.27% of Hindu Religion *(X^2 value= 30.32; p<0.01)*; 68.57% of the patients had Education up to graduate /post graduation *(X^2 value= 97.81; p < 0.01);* 45.83% of patients doing job *(X^2 value= 40.22; p < 0.01)*; 32.43% of the Social class II *(X^2 value= 15.31; p<0.01*) were found to had good knowledge and it was statistically significant.

DISCUSSION

The present study was carried out during in Janak Smarak hospital of Vyara. A total of 225 participants, 75 of which were patients of Sickle Cell Disease & the remaining 150 had Sickle Cell Trait were included in the study. It was observed that proportion of females affected was more than that of their male counterparts which could have been because a higher sex ratio of Tapi district (1004 females per thousand males as per census 2011)[8].A study conducted by ShresthaA et al [9] and Jain B et al [10] found that males were more affected than females.

Table 3: Percentage of sickle cell disease & sickle cell trait patient, who knew about Sickle Cell Anemia Control Program run by the government

Knowledge about the program	SCD (n=75) (%)	SCT (n=150) (%)	Total (n=225) (%)
Yes	63 (84)	141 (94)	204 (91)
No	12 (16)	9 (6)	21 (9)
Total	75 (100)	150 (100)	225 (100)

Majority of participants belong to Gamit caste, followed by Chaudhari caste because of a higher population of these communities residing in the district (Data of Census 2011) [8]. The present study revealed that marital status was significantly associated with occurrence of SCD & SCT status. This can lead to higher chances of future transmission of sickle cell in the progeny due to higher prevalence of consanguineous marriages in the tribal communities.

Table 4: Association between levels of knowledge among sickle cell patients with their Socio-demographic Profile of the study subjects

Variable	Good (>11) Number (%)	Not so good(< 11) Number (%)	Total Number (%)	Chi-square Test
Age *(Years)				
18-45	15(60)	114(89.76)	129(84.86)	12.185
46- 100	10 (40)	13(10.23)	23(15.13)	P < 0.01
Total	25	127	152	
Caste				
Gamit	25 (67.56)	131(69.68)	156(69.33)	9.024
Chaudhari	8 (21.62)	54 (28.72)	62(27.55)	P <0.05
Others #	4 (10.81)	3 (1.59)	7(3.11)	
Total	37	188	225	
Marital Status				
Married	21(58.33)	85(70.83)	106(67.94)	2.96
Unmarried	13(36.11)	33 (27.5)	46(29.48)	P<0.23
Widow/widower	2(5.55)	2(1.66)	4(2.56)	
Total	36	120	156	
Religion				
Hindu	26(70.27)	183 (97.34)	209(92.88)	30.321
Christian	11(29.72)	5 (2.65)	16(7.11)	P <0.01
Total	37	188	225	
Education**				
Illiterate	1 (2.85)	31(16.66)	32(14.47)	97.81
Primary (1-7 standard)	3(8.57)	62 (33.33)	65(29.41)	P <0.01
Secondary (8-10 standard)	2(5.71)	61 (32.79)	63(28.50)	
Higher Secondary(11-12 standard)	5 (14.28)	23 (12.36)	28(12.66)	
Graduate/PG/Diploma	24 (68.57)	9 (4.83)	33(14.93)	
Total	35	186	221	
Occupation***				
Student	6 (25)	98 (58.68)	104(54.45)	40.226
Unemployed	7 (29.16)	61 (36.52)	68(35.60)	P <0.01
Job	11 (45.83)	8 (4.79)	19(9.94)	
Total	24	167	191	
Social Class				
CLASS -I	16 (43.24)	57 (30.31)	73(32.44)	15.31
CLASS - II	12(32.43)	115 (61.17)	127(56.44)	P <0.01
CLASS - III	8(21.62)	13(6.91)	21(9.33)	
CLASS - IV	1 (2.70)	1 (0.53)	2(0.88)	
CLASS - V	0	2 (1.06)	2(0.88)	
Total	37	188	225	

"Others" include Valvi and Kokani castes.

* Age below the 18 years of age was excluded as they were not asked about the knowledge

** For Education, children less than 18 years of age (out of school**)** were excluded as they were not asked about the knowledge

*** For Occupation, children less than 18 years of age (out of school**)** were excluded as they were not asked about the knowledge

A study by M Kamble et al [11] also describes about history of consanguinity marriage; out of which (8.2%) had sickle cell disease and (5.2%) had sickle cell trait. It is hence extremely necessary to carry out genetic counselling after knowing sickle cell status among potential couples before marriage to avoid the further transmission in the successive generations.

In the present study only 11 % of the subjects were aware about hereditary cause of disease transmission. Also study by Gamit C et al [12] showed same result as 9% of study subject knew hereditary cause of disease. In contrast to this result, it was seen that study conducted by Coretta et al [13] had reported 45% of the patients and another study by Kofi et al[14] observed about 75% patients knew the hereditary nature of sickle cell disease.

In present study, 24 % of the subjects had knowledge regarding sign and symptoms. In the study conducted by Gamit C et al [12] also showed that, 16% patients also knew the symptoms of sickle cell disease and Coretta et al [13] also noted that 36% of the study subjects knew the sign & symptoms of Sickle cell disease. Opposite to this result, a study by Patil SS et al. [15] showed that amongst the study subjects, 75.86% had knowledge about symptoms of sickle cell disease and also whereas Kofi et al [14], reported 95% patients knew the symptoms of sickle cell disease. Among participants, only 6 % of the subjects knew their current haemoglobin status .The same result also seen by Gamit C et al [12] having only 2.5 % of study subject knew the haemoglobin status. A study in Nigeria among university students between the ages of 20 and 24 found that 86% knew about their haemoglobin status, and also only 18% of the subjects knew sickle cell crisis identification. For the awareness regarding nearest blood bank with respect to their residential location only 15 % of the subject knew about it.

Present study showed that out of 225 sickle cell patients, 91 % of the patients knew about sickle cell anaemia control program run by the government. A study by Gamit C [12] explained the need of strengthening of Sickle Cell Anaemia control program by improving in manpower resources especially true in relation to counsellors so that eventually the knowledge of Sickle Cell Status positive persons can be improved upon.

Out of the total 6 variables, total 3 variable like knowledge regarding disease transmission (Hereditary or not), sign & symptoms and current Haemoglobin status of the study participants were taken as "Primary Knowledge" because it is desirable for all the SCD and SCT individuals to know about them. The remaining 3 variable like Sickle Crisis Identification, Blood Transfusion in case of emergency and Nearest blood bank with respect to their residential location in case of Emergency were considered as "Core Knowledge" because these variables are not only must know aspects of the disease but they also play a critical role at the time of crisis to save lives of sufferers. In our study socio demographic variable like age group between 18-45 years, Gamit Caste, Hindu Religion, Education up to graduate /post graduation; patients doing job and Social class II showed association with having good knowledge. Also, study by Al-Suwaid et al [16] also showed that the association between the levels of knowledge of sickle cell anaemia patients with their socio-demographic characteristics like age group of 15to 30 , marriages, education up to secondary or diploma , unemployed, lower socio economical people having good knowledge and it was statistically significant.

CONCLUSION AND RECOMMENDATIONS

It can be concluded from the study that as the overall Knowledge regarding disease was averaging only at15 % which points out that the awareness seems to be grossly deficit even among the affected. It hence shows that the counseling of the disease among the affected was not adequate. This needs to be re-iterated in the IEC programmer especially in areas with high prevalence of Sickle cell disease and trait. IEC programme should have a dual pronged approach where the focus should be not only in hospitals for counseling the affected but also in the community to make them aware. Key areas of the disease should be explained to not only those affected but also the family members and the community in general. This will help in prevention of health situation like crisis and will help in prevention and restoration of health among the affected. Genetic counseling must be regularly carried out to ensure that the transmission of disease doesn't occur in future generations.

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