Original Research Article

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Knowledge, attitude and practice regarding sickle cell disease in adult sufferers and carriers in a rural area

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ABSTRACT

Background: According to State Health Society, there are about 30 lakhs sickle cell carrier and 1.5 lakhs sufferers in Maharashtra, prevalence being more in tribal population. As Sickle cell disease is associated with increased morbidity and mortality, it is important for patients to have awareness regarding its cause, treatment and genetic implication. To assess Knowledge, Attitude and Practices regarding sickle cell disease in adult sufferers and carriers.

Materials and Methods: The present study is an cross sectional study, carried out in 232 adult sickle cell disease sufferers and carriers in rural area of Chandrapur district from July 2014 to December 2015.

Results: Out of 232 study subjects, 17.24% were sufferers and 82.76% were carriers. Only 62.50% knew the correct cause and only 42.67% had correct knowledge of types of disease. 76.30% had correct knowledge about symptoms of disease. 40.09% patients were availing treatment of some kind. It was found that only 6.77% had done premarital screening of their spouses and 100% had screened their children for sickle cell disease.

Conclusion: Knowledge regarding sickle cell disease, its cause, types, treatment and prevention is satisfactory in sufferers but not in carriers. Overall attitude towards treatment and prevention is more or less positive. Treatment seeking behavior is good in sufferers but not in carriers.

Keywords: Sickle Cell disease, Knowledge, Attitude, Practice

INTRODUCTION

Sickle cell disease is an autosomal recessive, unifactorial or single gene haemoglobinopathy responsible for considerable morbidity and mortality. Sickle cell disease is expressed when sickled hemoglobin (HbS) is inherited from both parents i.e. sufferer (SS), while the heterozygous state (AS) is a carrier of disease called sickle cell trait. According to WHO, about 5% of world's population carries the gene responsible for hemoglobin disorders and about 300000 children are born worldwide with sickle cell disease every year. 4,5

In India, HbS was first detected in Veddoid tribe in Nilgiri hills of Tamilnadu, in 1952 by Lehman and

Cutbush.⁶ The incidence varies from 5% to 34% and it is mainly restricted to the tribal population.⁷ In India, it is the second most common haemoglobinopathy, next to Thalassemia.⁸ According to State Health Society; there are about 30 lakhs sickle cell carrier and 1.5 lakh sickle cell disease sufferers in Maharashtra. Out of these, about 10.5 lakh sickle cell carriers and 70 thousand sickle cell disease sufferers belong to the tribal population.^{9,10}

Sickle cell patient presents with anaemia, recurrent episodes of pain, recurrent infections, jaundice, stroke and delayed growth. Complications of sickle cell disease include serious infections, damage to vital organs, stroke, kidney damage, respiratory problems, bone marrow failure, growth failure, cognitive impairment,

maturational delay in children as well as high maternal and foetal morbidity and mortality. Recurrent complications interfere with the patient's life, especially with regard to education, work, and economic, social and psychosocial development. 11,12

Progress in the scientific knowledge of SCD and community awareness programmes have made it possible for patients to have access to treatment techniques that have helped many of them to live longer than before. However, misconceptions and misbeliefs still persists that affect the health seeking behaviour of the community

There is a need to assess underlying misconceptions because it will help to facilitate changes in service provision. Understanding the knowledge, attitudes and practices will help to fashion appropriate public health education programs to increase awareness and knowledge of the condition.

With this background, a study was carried out to assess knowledge, attitude and practices amongst sufferers and carriers of sickle cell disease in a rural area.

METHODS

This cross-sectional study was conducted in four villages of sub center Pombhurna, under Primary Health Centre, Pombhurna of Chandrapur district about 200 km from Nagpur, during July 2014 to December 2015.

Sample size of 232 was estimated from pilot study.

Approval from the Institutional Ethics Committee was sought. Permission from District Health Officer, Chandrapur was obtained.

One house in the village was selected randomly and every consecutive house was visited for sickle cell patients aged 18 years or more, till the sample size of 232 was achieved. Informed consent was taken from each study subject after explaining the purpose of the study.

Data was collected by interview method using structured questionnaire in the local language (Marathi). Information regarding socio-demographic characteristics like age, gender, marital status, and knowledge, attitude and practices regarding sickle cell disease was recorded in a pre-designed and pre-tested proforma.

Statistical analysis

Data was analysed using statistical software EPI Info 7. Descriptive statistics (percentage, mean, standard deviation, range) were used to summarise baseline characteristics of the study subjects. Fisher Exact test was used wherever required, p value < 0.05 was considered to be statistically significant.

RESULTS

Mean age of study participants was 41.9 ± 16.37 years with a range of 18-70 years. Out of 232 study subjects, 102(44%) were males and 130(56%) females. 55 (23.71%) were unmarried, 146 (62.93%) were married, 27(11.64%) widow/widower, 2(0.86%) were divorced and 2(0.86%) separated.

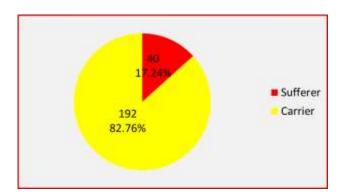


Figure 1: Sickle cell status of study subjects (n=232).

Out of 232 study subjects, 40 (17.24%) were sufferers and 192 (82.76%) were carriers.

Table 1: Knowledge	of study su	bjects regardii	ng sickle cell disease.
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	Study subjects having correct knowledge			
Knowledge of study subjects regarding sickle cell disease	Sufferer	Carrier	Total	
	No. (%)	No. (%)	No. (%)	
Carrier state of sickle cell disease	33 (82.50)	66(34.37)	99(42.67)	
Cause of sickle cell disease	38 (95.00)	107 (55.72)	145(62.50)	
Symptoms of sickle cell disease	40(100.00)	136(70.83)	176(75.86)	
Investigations for diagnosis of sickle cell disease	40 (100.00)	156 (81.25)	196(84.48)	
Knowledge regarding sickle cell crisis	14(35.00)	19 (9.89)	33(14.24)	
Precautions to be taken to live healthy life	38 (95.00)	129 (67.18)	167(71.98)	
Treatment to be taken for sickle cell disease	40 (100.00)	99 (51.56)	139(59.91)	
Knowledge regarding transmission of sickle cell disease	37(92.50)	89(46.35)	126(54.31)	
Knowledge regarding prevention of sickle cell disease	30(75.00)	90(46.87)	120(51.72)	

Amongst the study subjects 33(82.50%) sufferers and only 66 (34.37%) carriers had correct knowledge regarding the carrier state of sickle cell disease. Only 145 (62.50%) study subjects had knowledge regarding hereditary nature of sickle cell disease. However, 02(5%) sufferers and 85(44.28%) carriers did not know the cause of sickle cell disease. The difference between the knowledge of the sufferers and carriers was found to be statistically significant (p=0.001, fisher exact test).

All the sufferers and 70.83% carriers had knowledge about symptoms of sickle cell disease like joint pain and yellowness of eyes, easy fatigability, pain in abdomen, fever, difficulty in breathing, chest pain and headache. 196(84.48%) study subjects knew that the investigation done for diagnosis of sickle cell disease is blood

examination. The difference between the knowledge amongst sufferers and carriers was found to be statistically significant (p=0.000, fisher exact test). Only 14.24% study subjects, had knowledge about sickle cell crisis, out of which 35% were sufferers and 9.89% carriers.

Amongst the lifestyle measures to be practiced majority 167(71.98%) study subjects knew that exertion should be avoided, 102(43.96%) knew that daily intake of 8-10 glasses of water should be taken, 56(24.13%) knew that green vegetables should be consumed, and a few had knowledge that high altitude should be avoided by sickle cell patients to live a healthy life. 32.82% carriers were not aware regarding precautions to be taken to live healthy life for sickle cell disease.

Table 2: Attitude of study subjects regarding sickle cell disease (n=232).

A 44444 . J	Responses of study subjects				
Attitude	Strongly disagree	Disagree	Undecided	Agree	Strongly agree
Sickle cell disease is a curse of god	120	16	17	43	36
	(51.72)	(6.89)	(7.32)	(18.53)	(15.54)
Only poor people suffer from sickle cell disease	130	08	14	38	42
	(56.03)	(3.44)	(6.03)	(16.37)	(18.13)
Sickle cell disease being a genetically transmitted disorder, not much can be done to prevent it	19	15	135	32	31
	(8.18)	(6.46)	(58.18)	(13.79)	(13.39)
Health check-up should be done regularly for sickle cell disease	03(1.29)	05(2.15)	80(34.48)	32(13.79)	112(48.29)
There is a need to take regular treatment for sickle cell disease	09(3.87)	07(3.01)	89(38.36)	25(10.77)	122(52.59)
Repeated blood transfusions can cure sickle cell disease	57(24.56)	22(9.48)	131(56.46)	12(5.17)	10(4.33)
There is need for pre-marital screening of spouse for sickle cell disease	05	13	170	23	21
	(2.15)	(5.60)	(73.27)	(9.91)	(9.07)
Pre-natal diagnosis is necessary to prevent sickle cell disease	13	18	165	16	20
	(5.60)	(7.75)	(71.12)	(6.89)	(8.64)
All children should be screened for sickle cell disease	00	03	28	24	177
	(0.00)	(1.29)	(12.06)	(10.36)	(76.29)
Sickle cell disease as an illness is less known in society and needs more awareness	00 (0.00)	00 (0.00)	56 (24.13)	26 (11.20)	150 (64.67)

Amongst the study subjects, 139(59.91%) had knowledge regarding medicines to be taken by sickle cell patients while 90(48.44%) carriers did not know regarding them.

Only 54.31% study subjects had correct knowledge regarding transmission of sickle cell disease. 120(51.72%) study subjects knew that sickle cell disease can be prevented. Overall knowledge regarding transmission and prevention of sickle cell disease was poor in carriers as compared to sufferers.

When the attitude of the study subjects was assessed more than half of the study subjects disagreed that sickle cell disease is a curse of god and only poor people suffer from sickle cell disease. Amongst the study subjects, 58.18% were undecided regarding sickle cell disease being a genetically transmitted disorder, and felt not much can be done to prevent it. Around 60% of the study subjects agreed that health check-up should be done regularly and there is need to take regular treatment for sickle cell disease.

Amongst the study subjects 56.46%, were undecided regarding repeated blood transfusions can cure sickle cell disease and 34.04% disagreed. Only 18.98% agreed about the need for pre-marital screening of spouse for sickle cell disease while 15.53% study subjects agreed regarding the necessity of pre-natal diagnosis for

prevention of sickle cell disease. Majority of the study subjects agreed that all children should be screened for sickle cell disease and sickle cell disease as an illness is less known in society and needs more awareness.

Only 89(38.36%) study subjects were undergoing regular health check-up for sickle cell disease. All the sufferers

and only 25.52% of carriers did regular health check-up for sickle cell disease.

Amongst the study subjects 85(36.63%) were taking treatment out of which 40(100%) were sufferers and only 45(23.43%) carriers.

Table 3: Practices of study subjects regarding sickle cell disease.

	Study subjects				
Practices	Sufferer	Carrier	Total		
	No. (%)	No. (%)	No. (%)		
Regular health check-up for sickle cell disease	40(100.00)	49(25.52)	89(38.36)		
Treatment for sickle cell disease	40(100.00)	45(23.43)	85(36.63)		
Precautions taken to live healthy life	35(87.50)	79 (41.14)	144(62.06)		
Practices of married study subjects regarding screening for	Sufferer	Carrier	Total		
sickle cell disease	N=25	N=152	N=177		
Done pre-marital screening of spouse for sickle cell status	07(28.00)	05(3.28)	12(6.77)		
Spouse screened after knowing their positive sickle cell status	25(100.00)	113(74.34)	138(77.96)		
Children screened for sickle cell disease	25(100.00)	152(100.00)	177(100.00)		

Majority of the study subjects avoided exertion; some drank 8-10 glasses of water daily, while a few consumed green vegetables and avoided high altitude. Practices of study subjects regarding precautions taken to live healthy life were good in sufferers but not of acceptable level in carriers.

Only 12 (6.77%) study subjects had done pre-marital screening of their spouses for sickle cell status. Amongst them 07 (28%) were sufferers and 05 (3.28%) were carriers. 138(77.96%) study subjects had got their spouses screened after knowing their positive sickle cell status. Amongst them 25 (100%) were sufferers and 113 (74.34%) were carriers. All study subjects i.e. 25 (100%) sufferers and 152 (100%) carriers had screened their children for sickle cell disease.

DISCUSSION

In the year 2006, Department of Health & Family Welfare, Government of India launched sickle Cell disease control program. Under this Programme, mass sickle cell screening was taken up by Government of Maharashtra state. This study is an endeavour to reach people of the rural areas, in the age group 18 years or above, having an abnormal sickle cell gene, and are either sufferers or carriers of sickle cell disease.

Out of the 232 study subjects, 40 (17.24%) were sufferers and 192 (82.76%) were carriers. In the present study number of sufferers were more compared to the study conducted by Gamit et al who reported, 96% carriers and 4% sufferers. In the present study, 62.50% study subjects had knowledge regarding the hereditary nature and not even half of them were aware of the carrier state of sickle cell disease. Gamit et al, found only 9% study subjects knew the hereditary nature of sickle cell disease whereas Coretta et al, reported 45% and Kofi et al

observed that 75% patients knew the hereditary nature of sickle cell disease. ¹³⁻¹⁵

Amongst the study subjects, 75.86% had knowledge about symptoms of sickle cell disease like joint pain and yellowness of eyes, pain in abdomen, etc. Sufferers had better knowledge than carriers regarding symptoms of sickle cell disease. In the study conducted by Gamit et al, 16% patients knew the symptoms of sickle cell disease. Coretta et al, noted 36% whereas Kofi et al, reported 95% patients knew the symptoms of sickle cell disease. ¹³⁻¹⁵ Only 14.24% study subjects out of which 35% sufferers and 9.89% carriers had knowledge about sickle cell crisis.

In the present study, 121(68.34%) study subjects were aware about sickle cell status of their spouses. In the study conducted by Gamit et al, only 17% patients knew the sickle cell status of their family members. Treadwell et al, reported 12% and Nazir et al, found that 36% patients knew the sickle cell status of their family members. In the present study, high proportion of study subjects was aware about sickle cell status as entire population of this area was screened for sickle cell disease.

All married sufferers were aware about sickle cell status of their spouses. However, 36.84% married carriers were not aware about sickle cell status of their spouses. Spouses of 9.21% carriers had expired and spouses of 27.63% carriers were tested but they were not aware about their SCD status.

Sufferers had fair knowledge than carriers regarding precautions to be taken by them to live healthy life like avoiding exertion, daily intake of 8-10 glasses of water, consumption of green leafy vegetables, and avoiding high altitude. This might be as more efforts were focused on

sufferers since the launching of the sickle cell disease control program.

In this study, 59.91% study subjects knew that folic acid should be taken regularly for sickle cell disease. Coretta M et al and Treadwell M et al, reported lower proportion of knowledge compared to the present study. ^{14,16} Kofi et al (2010) and Nazir et al (2005), reported higher proportion of knowledge regarding regular intake of folic acid compared to our study. ^{15,17}

In the present study, 54.31% study subjects had knowledge regarding transmission of disease. 51.72% study subjects knew regarding its prevention. Overall knowledge regarding transmission and prevention of sickle cell disease was poor in carriers as compared to sufferers. Lack of correct knowledge about sickle cell disease will prevent this tribal high risk population to come to health care facility even the services are available.

Attitude of the study subjects regarding sickle cell disease was assessed. It was observed that, 136(58.61%) study subjects disagreed that sickle cell disease is a curse of god. Almost half of the study subjects disagreed that only poor people suffer from sickle cell disease.

In this study, majority of the study subjects were undecided regarding sickle cell disease being a genetically transmitted disorder, and felt not much can be done to prevent it.

In the present study 144(62.08%) study subjects agreed that there is need to take regularly treatment for sickle cell disease. Similar findings were reported by Olatano et al, Coretta et al and Kofi et al. 14,15,18

Few study subjects (9.50%) agreed that repeated blood transfusions can cure sickle cell disease. Coretta M et al observed that 19% patients and Nazir et al reported that 49% patients believed that sickle cell disease is curable with repeated blood transfusions. Amongst the study subjects, about 20% agreed that there is need for premarital screening of spouse for sickle cell disease. Similar findings were noted by Gamit C et al. Treadwell et al and Nazir et al. 16,17 Attitude towards pre-marital screening of spouse for sickle cell disease was not positive.

Only 36(15.53%) study subjects agreed that pre-natal diagnosis is necessary to prevent sickle cell disease. In the study conducted by Coretta et al(2010), 29% sickle cell patients agreed that physician opinion regarding prenatal diagnosis was must before planning for future pregnancy. Majority of the study subjects agreed that all children should be screened for sickle cell disease and 75.87% study subjects agreed that sickle cell disease as an illness is less known in society and needs more awareness.

Practices regarding sickle cell disease revealed that, 38.36% study subjects were undergoing regular health check-up for sickle cell disease. Similar findings were reported by Gamit et al However Coretta et al, Kofi et al and Nazir et al, observed that majority of patients were undergoing regular health check-up for sickle cell disease. ^{13,14,15,17}

Amongst the study subjects undergoing regular health check-up, 100% were sufferers and only 25.52% were carriers. This shows that practices regarding regular health check-up for sickle cell disease were not of acceptable level in carriers.

In the present study, 36.63% study subjects were taking treatment. This is in accordance with the findings of Gamit et al, Coretta et al and Treadwell et al. ^{13,15,17} Practices of study subjects regarding precautions taken to live healthy life were good in sufferers but not of acceptable level in carriers. Only 28% sufferers and 3.28% carriers had done pre-marital screening of their spouses for sickle cell status. 77.96% study subjects had got their spouses screened after knowing their positive sickle cell status. Children of all study subjects were screened for sickle cell disease.

This study shows that knowledge regarding cause, types of sickle cell disease, sickle cell crisis, precautions to be taken to live healthy life, treatment and prevention of sickle cell disease is satisfactory in sufferers but not of acceptable level in carriers. Overall attitude of the sufferers as well as carriers towards treatment and prevention of sickle cell disease is more or less positive.

All sufferers are taking treatment, however almost half of the carriers are not taking treatment of any kind. Practices related to pre-marital screening of spouse and pre-natal diagnosis of sickle cell disease is infrequent in both sufferers and carriers.

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