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Original Research Paper A Community Based Sickle Cell Screening Program in Pilgrims Using Camp Approach

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Abstract

Background: Sickle cell anemia is a common genetic condition due to haemoglobin disorder-inheritance of mutant haemoglobin genes from both or either parents. The carrier frequency of haemoglobinopathy varies between 3 and 17% in different populations of India. Though the prevalence is high, limited studies are available on this topic. As prevention is only solution, there is a need to screen the general population on mass basis.

Objectives: To study the prevalence of Sickle cell anemia among pilgrims in central India and to create awareness regarding Sickle cell anemia in general population.

Material and Method: The present cross sectional study was carried out at camps at Dikshabhoomi and Durgamata mandir, Nagpur in first week of October 2015; among visitors coming there. Health education regarding Sickle cell was given to them in small groups. Detailed clinical history was taken, Body Mass Index was calculated and intravenous blood samples were taken. Solubility test was done for Sickle cell anaemia and positive test samples were processed for electrophoresis for AS or AA type.

Result: Prevalence of Sickle cell anemia was 6.25%, common in males and 11-30 years age group. **Conclusion**: There is a need of mass screening for Sickle cell disease in general population from all the age groups and all the caste to prevent the spread and complications of Sickle cell disease. **Keywords:** Sickle cell anemia, Solubility test, Caste, Hemoglobin disorder.

Introduction

Sickle cell anemia (also known as Sickle cell disorder or Sickle cell disease) is a common genetic condition due to hemoglobin disorderinheritance of mutant haemoglobin genes from both or either parents. Such haemoglobinopathies, mainly thalassemias and Sickle cell anaemia, are globally widespread. About 5% of world's population carries genes responsible for haemoglobinopathies. About 7% of the world's population is carriers of some form of hemoglobin disorder. There are about 270 million carriers of Sickle cell anemia and or thalassemia¹.

With a population of over 1.2 billion individuals, it is estimated that India is home to over 50% of the world's SCD patients. While Sickle cell is prevalent in many ethnic groups, the highest prevalence appears to be in three particular sociodisadvantaged economically communities, identified as the Scheduled Castes (SC). Scheduled Tribes (ST), and Other Backward Classes (OBC). Together these three groups comprise over 75% of the Indian population. The state of Maharashtra in Central India has a population of 112.3 million of which the SC and ST comprise 10.2% and 8.9% of the population respectively. Additionally, the OBC population comprises 52% of the population².

Sickle cell anaemia is particularly common among people whose ancestors come from Sub-Saharan Africa, India, Saudi Arabia and Mediterarian countries.

Sickle cell anemia covers a wide spectrum of illness. Most affected people have chronic anaemia with a haemoglobin concentration of around 8 mg/dl. Individuals with single globin chain defect are called carriers or traits or heterozygotes and they are generally for symptomless. When the gene sickle hemoglobin is inherited from both parents, it is called sickle cell anemia¹.

The population screening program uncovered previously undiagnosed cases, and provided detailed information for population based disease counseling. prevention programs and comprehensive care programs². This study was carried out with the objective of detecting Sickle cell anaemia in study subjects coming to the camp from all the castes and giving health education to them, so as to prevent the complications related to Sickle cell anaemia. Through this study. awareness is created among people about the disease. As prevention is the only solution at present, it is necessary to carry out such camps.

Material and Method

The present cross-sectional study was carried out at Dikshabhoomi and Durgamata mandir, Nagpur in Navaratra in first week of October 2015. Every year Dhammachakra Parivartan Day is celebrated at Dikshabhoomi, Nagpur and Navaratra is also celebrated at Durgamata mandir, Dhantoli, Nagpur at the same time. People from different places of Maharashtra and nearby places of other states from all the communities come to Dikshabhoomi and also visit Durgamata mandir. Camp was organized for Sickle cell testing at both the places by the Department of Preventive and Social Medicine and Regional Sickle cell unit, Department of Pathology, Indira Gandhi Government Medical College, Nagpur. Permission from ethical committee of Indira Gandhi Government Medical College, Nagpur was taken. Counseling of the persons visiting at one of the above camps was done for Sickle cell testing. Explanations were given regarding hereditary nature of the disease and the disease can be prevented but no curative treatment is available. Sample of convenience was included in the study. Total 1550 study subjects were enrolled in this study. Detailed clinical history of study subjects who were willing for the Sickle cell disease testing was taken. Body Mass Index (BMI) was calculated by measuring height and weight of all the study subjects.

Intravenous blood samples were taken for sickling tests. The blood samples were collected by trained personnel after taking informed consent of study subjects. The study subjects were screened by Dithionite tube turbidity (DTT) also known as solubility test in the present study as it is a rapid method and easy to be carried out in the field setting. The test is recommended by ICMR network on Sickle Cell Disorders coordinated by Institute of Immunohematology, Mumbai. The samples of those found positive for solubility test were processed in Department of Pathology for electrophoresis to find out Sickle cell trait (AS) as well as Sickle cell disease (SS); pattern of Sickle cell anaemia.

All study subjects who were having AS or SS disease were communicated by post about their disease and health education pamphlet was also

sent along with it. They were advised in writing to meet the medical officer nearby along with their reports.

Statistical analysis was done by statistical software Epi Info version 7. P value less than 0.05 was considered as statistically significant.

Results

(**Table No. 1**) Out of total 1550 study subjects screened, maximum i.e. 63.9 % were male. 35.2% of study subjects were in the age group of 21-30 years.

(**Table No. 2**) In the present study; the prevalence of Sickle cell anemia was 6.25% as 97 out of 1550 study subjects were having positive Sickle cell test. Sickle cell trait was seen in 6.06% (94) whereas Sickle cell disease was found in 0.19% (03) study subjects. Sickle cell anaemia was more common in males than females in the present study. Out of positive study subjects, 58.76% were males while 41.23% were females. 42.3% of subjects with positive sickling test were in the age group of 21 to 30 years.

AS-94, SS-03 (x²= 0.1613,P= 0.9225,df=2)

(Table No. 3) The study subjects with Sickle cell anaemia were mainly from Scheduled caste (79.4%) followed by Other backward classes (Teli, Kunbi, Sonar, Sutar, Banjara) and Scheduled tribe as well as other caste as shown in Table3. (x^2 = 34.96,P=0.0000001,df=1)

The association of Sickle cell anaemia and Brahmin caste was statistically significant i.e. the prevalence was less in Brahmin caste (P 0.02). Whereas the association of Sickle cell anaemia was statistically very highly significant in Scheduled caste (P0.0000001) and other caste (P0.00002).

Out of total 1550 subjects 716 (46.1 %) were not having any complaints, and 834 (53.9 %) were having one or more complaint. The most common complaint was generalized weakness in 613(73.5%), followed by other complaints such as generalized body ache, joint pain, fever, cough and cold, abdominal pain etc, 221(26.49%).

 $(x^2 = 0.3567, P=0.9490, df=1)$

(**Table No. 4**) When body mass index (BMI) was studied, total 67 (69.07%) subjects were of normal weight, 23(23.71%) subjects were undernourished and 7(7.21%) were overweight. There was no statistically significant association found between BMI and SCA.

(X²=5.327, p=0.06970, df=2)

Table 1: Distribution of Study Subjects according	
to age and sex	

Age(years)	Male	Female	Total (%)
1-10	65	45	110(7.1)
11-20	208	135	343(22.1)
21-30	357	189	546(35.2)
31-40	205	115	320(24.7)
41 & above	155	76	231(14.9)
Total	990(63.9)	560(36.1)	1550 (100)

Table 2: Age and Sex v	wise distribution of Sickle
cell anemia	

Age (years)	Male	Female	Total (%)	
1-10	03	04	07(7.2)	
11-20	16	08	24(24.7)	
21-30	24	17	41(42.3)	
31-40	07	07	14(14.5)	
41 & above	07	04	11(11.3)	
Total	57	40	97(100.0)	
$(x^2 = 0.1613, P = 0.9225, df = 2)$				

Table 3: Caste wise distribution of study Subjects

Caste	Study Subjects	Sickle cell anemia
	No. (%)	No. (%)
Scheduled caste	682(44.0)	77(79.4)
Scheduled tribes	74(4.8)	07(7.2)
OBC	444(28.7)	10(10.3)
Brahmin	89(5.7)	00(0.0)
Other	261(16.8)	03(3.1)
Total	1550 (100)	97 (100)

 $(x^2 = 34.96, P=0.0000001, df=1)$

Table 4: Distribution of Study Subjects according
to Body Mass Index (BMI)

BMI	Sickle cell anemia N=97(%)	Normal N=1453 (%)	Total N =1550 (%)	
Underweight	23 (23.71)	475 (32.69)	498(32.12)	
Normal weight	67 (69.07)	830 (57.12)	897(57.87)	
Over weight	07 (7.21)	144 (9.91)	151(9.74)	
Obese	00 (00)	04 (00.27)	04(00.34)	
$(\mathbf{y}^2 - 5327p - 006070df - 2)$				

(X²=5.327, p=0.06970, df=2)

Discussion

Prevalence of Sickle cell disease was 6.25% among study subjects in the present study. Ndeezi G. et al $(2016)^3$ carried out study in Uganda found

that the overall number of children with Sickle cell trait was 12 979 (13.3%) and with disease was 716 (0.7%). Deore A. U. et al, $(2013)^4$ conducted a study in Nagpur district of Maharashtra found the prevalence of Sickle cell anemia was 4.94%. However higher prevalence was noted by Dhumne UL et al. $(2011)^5$ as 18.3% in rural areas of Chandrapur, Maharashtra. Whereas Dangi CBS et al. (2010)⁶ mentioned 7% prevalence in randomly selected areas of Bhopal district and Kohne E et al, $(2010)^7$ observed 6.2% suffering from Sickle Cell Disorder at Ulm university, Germany. In a study conducted by Mandot S. et $al(2008)^8$, in Garasia Tribes of Rajasthan, the prevalence of Sickle cell anemia was found to be 9.2%. But lower prevalence of Sickle cell disorder was noted by Deshmukh P et al., (2006)⁹ as 2.9% in study subjects from rural Wardha. Sahu T et al, (2003)¹⁹ found 16.33% children sickling positive in tribal area of Gajapati district in South Orisa. Kamble M et al, (2000)¹⁰ found 5.7% prevalence in rural hospital of central India.

Sickle cell anaemia was more common in males in the present study. Doshi Neena et al, $(2011)^{11}$ observed the prevalence of Sickle cell disorder was equal in both the sexes. Higher prevalence in females was noted by Dhumne UL et al $(2011)^5$ as 8.5% prevalence in males and 9.8% prevalence in females. Similarly Deshmukh P et al, $(2006)^9$ found Sickle cell disease in 2.8% males and 3.0% females. Mukherjee MB et al, $(2004)^{12}$ noted that no significant differences were observed between the male and female children with sickle cell disease. Kamble M et al, $(2000)^{10}$ mentioned male: female ratio was 1.65:1 in HbSS cases and 1.71:1 in HbAS cases.

The study subjects were mainly from 21-40 years age group and Sickle cell disorder was common in 11-30 years males in the present study. This shows young males were the main group of visitors at above mentioned places. Agewise, Doshi Neena et al $(2011)^{11}$ mentioned 32.8 years as the mean age amongst the positive cases. Patra PK(2011)¹³ observed prevalence of Sickle cell

trait increased significantly with age but no age related trend occurred with SS disease. While Deshmukh P et al, (2006)⁹ observed Sickle cell disease mainly in 46-55 years and it increased with increasing age.

Sickle cell trait was common as compared to Sickle cell disease in the present study. Also Chandrashekhar V et $al_{(2011)}^{14}$ found 5.3% had HbS trait and 1 .4% had Sickle cell disease in study subjects at Chennai , Tamil Nadu. Likewise Doshi Neena et al (2011)¹¹, observed Sickle cell trait in 7.86% in one of the rural area of Gujarat. However Patra PK et al (2011)¹³, noted Sickle cell trait in 9.3% and Sickle cell disease in 0.21% children aged 3-15 years in rural area of Raipur, Chhatisgarh. Similarly Adly G et al, (2008)¹⁵ observed prevalence of Sickle cell trait in 7.5% and Sickle cell disease in 0.46% patients at Khaula hospital ,Oman. Balgir RS(2005)²⁰ found prevalence of Sickle cell trait in 29.8% and Sickle cell disease in7.5%.

In the present study, study subjects were mainly from Scheduled caste community and Sickle cell disease was commonly observed in Scheduled caste followed by Kunbi, Teli, Gond , NT and other. This finding suggests need of Sickle cell testing in people from all the caste.

Deore A.U. et al, $(2013)^4$ conducted a study in Nagpur district of Maharashtra found the highest frequency of the disease was observed in Gond (14.28%) followed by Pradhan (11.49%), Bhil (11.42), Teli (10.48%), Matang (8.75%) and Korku (7.89%) whereas lowest frequency was recorded from Muslim Gawali, Bhangi and Koli Mahadeo. Patra PK et al (2011)¹³, mentioned highest frequency in SC, ST and OBC which ranges from 10% to 24%. Gupta RB $(2006)^{16}$ mentioned that the problem of Sickle cell gene in Madhya Pradesh exist among Scheduled caste, Scheduled tribes and backward class communities. Colah R.B. et al $(2015)^{18}$ the prevalence of Sickle cell carriers in different tribes varies from 0 to 35 per cent. The tribal groups with a high prevalence of HbS (20-35 %) include the Bhils, Madias, Pawaras, Pardhans and Otkars. It has also been

estimated that Gadchiroli, Chandrapur, Nagpur, Bhandara, Yoetmal and Nandurbar districts would have more than 5000 cases of Sickle cell anaemia. Kamble M et al, (2000)¹⁰, mentioned Sickle cell disease was maximum in Mahar community (70%) followed by Kunbi (8%) and Teli (6%).However Kate SL et al, (2002)²¹ observed the overall prevalence amongst SC, ST and OBC was 10%.

Fifty –Ninth World Health Assembly (2006)¹⁷ mentioned that the manifestations of Sickle cell anemia are more unpredictable and variable than those of thalassemia's. Many affected individuals, however, have a good quality of life, and in some parts of the world (Bahrain, India, Eastern Saudi Arabia) additional genetic factors may reduce the severity of the disease. Similarly in the present study the association of presenting complaints with Sickle cell anaemia was statistically non significant.

Conclusions

Prevalence of Sickle cell disease was found to be 6.25% in the present study. Prevalence of Sickle cell anaemia was more common in scheduled caste community but it is seen in other caste also. Thus there is a need of screening for Sickle cell disease in general population to prevent the spread and complications of Sickle cell disease.

Limitations

Since it is a camp based study and the study population is not a true representative of the general population, the results cannot be generalized. Sampling procedure and sample size was mainly based on camp approach.

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Conflicts of interest

There are no conflicts of interest.

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