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Prevalence of Sickle Cell Anemic Subjects from Gadchiroli District, Maharashtra, India

S. R. Kohchale[1], Misar S. D.[2]
[1]*Shri R.L.T. College of Science, Akola, M.S., India*
[2]*Janata Mahavidyalaya Chandrapur M.S., India*

Abstract: Sickle cell disease is caused by Mutations in the HBB gene. Hemoglobin consists of four protein subunits, typically, two subunits called alpha-globin and two subunits called beta-globin.
People with sickle cell anemia suffers with high morbidity and with many intercurrent infections, people of study district are with high economic burden, terminate fatality in childhood state and have the emotional and psychological trauma including the family members the exact magnitude of the problem in the study district is still obscure.
The study conducted from April 2009 to April 2012 to know the prevalence of sickle cell anemia by month long survey and by visiting all PHC'S and RH of district and data collected to know the prevalence of sickle cell trait and sickle cell disease total 7763 cases were recorded in present study and age wise, gender wise and caste wise distribution recorded and the data was analyzed statistically.
Keywords: Sickle, anemia, Gadchiroli, Haemoglobin, beta-globin

I. INTRODUCTION

Sickle cell anemia is known to the medical world since the discovery of this entity by Dr. James Herrick, (1910) a Chicago cardiologist. The highest frequency of causative gene of sickle cell anemia is in tropical Africa it occurs with lower frequency in Mediterranean basin, Saudi Arabia and parts of India. In India high frequencies of sickle cell gene is reported from states of Orissa, Maharashtra, Madhya Pradesh, Gujarat, and parts of Kerala (Kaur *et al.*, 1997, Shah 2004, Chhotray *et al.*, 2004). In central India the maximum number of cases of sickle cell anemia is seen in Mahar, Gond, Teli, and Kunbi. It is estimated the sickle cell gene is prevalent in 75 districts (in various states) in India. Before this century most of the individual with sickle cell anemia died before the reproductive age. Sickle cell trait has its highest prevalence in areas endemic for malaria suggested that Hb-S offered selective removal of sickled from the circulation probably reduces degree of parasitemia and substantially limits the infection process. In India first case of sickle cell anemia was reported by Lehman and Cutbush, 1952 and later by Jain *et al.*, 1981 and thereafter awareness of this was increased. Various aspects of this disease including peculiar prepaundance in few communities have been studied. In India this condition is common among certain tribes in south India, Asam, Bihar, and Orissa. In Maharashtra it is reported in specific communities as well as community from Gujarat (Shah, 2004). The incidence of sickle cell gene is high in communities like Mahar, Kunbi as reported by Shukla and Solanki, (1958) incidence of sickle cell anemia in black population is also high.

II. MATERIAL AND METHODS

The study conducted in Gadchiroli district, located Latitude 20^{o} 10' 56.66"N and Altitude 80^{o} 0' 11.46" E, of Maharashtra with population of 9 to10 lakh where most of the area is covered by dense forest having a major population of Gond and Madia tribes.The study conducted from April 2009 to April 2012 to know the prevalence of sickle cell anemia by month long survey and by visiting all PHC'S and RH of district and data collected to know the prevalence of sickle cell trait and sickle cell disease total 7763 cases were recorded in present study and the data was analyzed statistically.

III. OBSERVATION AND RESULTS

A. Age Wise Distribution

Data was collected and analyzed statistically, in the present study total Hb-AS were found to be 6638 (85.51%), and 1125 (14.49%) Hb-SS of total affected population. In patients with Hb-AS (carrier) in age group of 0 to 10 years were 1460 (21.99%) and in Hb-SS (sufferers) were found to be 333 (29.6%). In age group of 11 to 15 in Hb-AS were 1279 (19.26%) and in Hb-SS were 201(17.86%). Between age group of 16-20 year Hb-AS were 1075 (16.19%) and Hb-SS were 194 (17.24%). In age group of 20 years above patients in Hb-AS were 2824 (42.54%) and in Hb-SS were 397 (35.28%) **(Table - 1, Fig-1).**

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B. Gender Wise Distribution

In 0-10 year population Hb-AS male were 727(10.95%) females 733 (11.04%), in Hb-SS males 169 (15.02%) females 164 (14.57%). In age group of 11-15 in Hb-AS patients male were 535 (8.05%) females were 744 (11.20%) and in Hb-SS males 83 (7.37%) females 118 (14.57%). In Hb-AS patients in the age group of 16-20, 331 male and 744 female (4.98% male and 11.20% females) out of them 23 were married and 308 were unmarried (0.34% and 4.64%), in Hb-SS males 79 (7.02%) married 5 (0.44%) , unmarried 74 (6.57%) and 115 (10.22%) females, 19 (1.68%) married and 96 (8.53%) unmarried. Above age group of 20 years in Hb-AS 834 (12.56%) were male 687 (10.34%) were married and 147 (2.21%) unmarried and 1990 female (29.97%) out of them 1835 (27.64%) were married and 155 (2.35%) were unmarried. In Hb-SS above 20 years age group male were 153 (13.6%) 108 (9.6%) married and 45 (4%) unmarried, female were 244 (21.68%) married 217 (19.28%) and 27 (2.4%) unmarried were found in the total affected population (Table- 1, Fig-1)

Table: 1 Showing Age and Gender wise distribution

	Total no. of patients				Total
	Male		Female		
	Hb-AS	Hb-SS	Hb-AS	Hb-SS	
0-10 years	727 (10.95%)	169 (15.02%)	733 (11.04%)	164 (14.57%)	1460 (21.99%)
11-15 years	535 (8.05%)	83 (7.37%)	744 (11.20%)	118 (14.57%)	1279 (19.26%)
16-20 Years	331 (4.98%)	79 (0.02%)	744 (11.20%)	115 (10.22%)	1075 (16.19%)
20 Years above	834 (12.56%)	153 (13.6%)	1990 (29.97%)	244 (21.68)	2824 (42.54%)

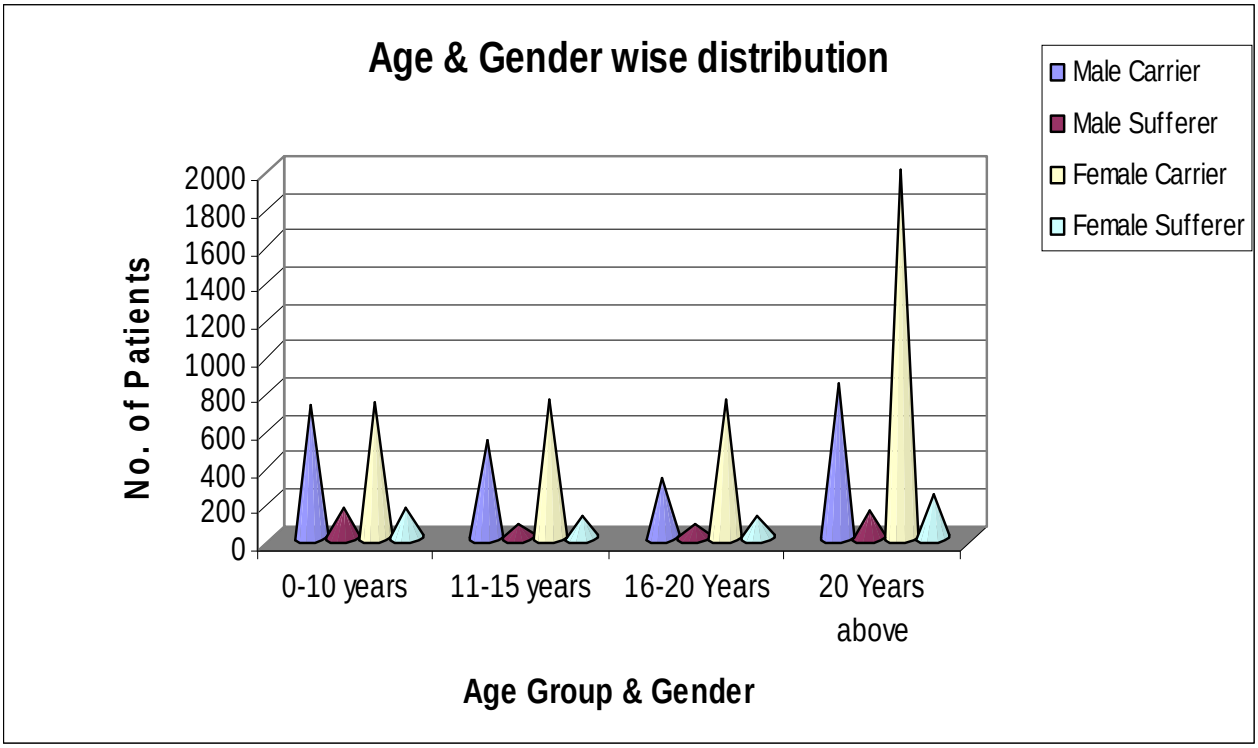


Fig. 1: Showing Age & Gender wise distribution

C. Caste Wise Distribution

In present study total Hb-AS were 6638 and total Hb-SS 1125, total affected population in schedule caste were 2754 (35.47%) out of them 2266 (34.13%) were Hb-AS and 488 (43.37%) were Hb-SS, among Scheduled tribe with maximum patients 2854 (36.76%) with 2418 (36.43%) Hb-AS and 436 (38.75%) were Hb-SS. In other population total other were 2155 (27.75%) out of which 1954 (29.43%) were Hb-AS and 201(17.81%) were Hb-SS **(Table-2 and Fig -2).**

Table: 2 Showing Cast wise distribution

SC		ST		OTHERS	
Hb-AS	Hb-SS	Hb-AS	Hb-SS	Hb-AS	Hb-SS
2266 (34.13%)	488 (43.37%)	2418 (36.43%)	436 (38.75%)	1954 (29.43%)	201 (17.81%)

Total SC-2754 (35.47%), TOTAL ST-2854 (36.76%), Total Others-2155 (27.75%)

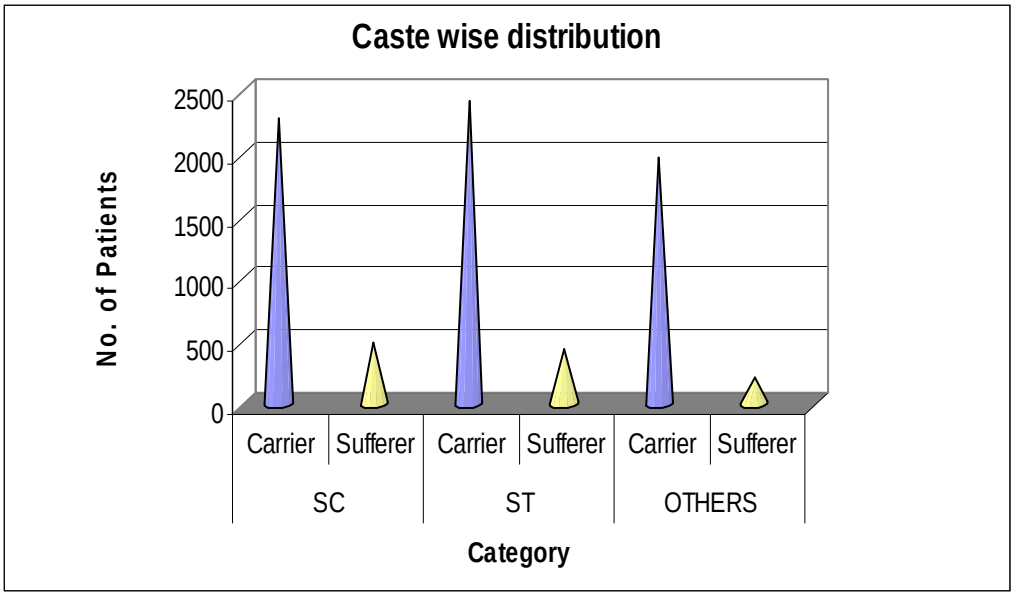


Fig. 2: Showing Caste wise distribution

IV. DISCUSSION

A. Distribution with Age, Gender and Caste

All the screened patients were analyzed according to age, gender and caste and finally their distribution was calculated statistically. The age wise prevalence of sickle cell anemia patients in Gadchiroli districts, reported as 21.99% in age group of 0 to 10 years were carriers with Hb-AS and 29.6% were Hb-SS (sufferers) In age group of 11 to 15 in Hb-AS were 19.26% and in sufferers were 17.86%. Between age group of 16-20 year Hb-AS carriers were 16.19% and Hb-SS sufferers were (17.24%). In age group of 20 years above patients in Hb-AS were 42.54% and in Hb-SS sufferers were 35.28%.Thus the maximum Hb-AS carrier and Hb-SS sufferer patients were above 20 years of age. This finding is in accordance with Leikin *et al.*, (1989) who also found that the patients of sickle cell disease were distributed more in patients of higher age groups. Ankushe (1993) and Kamble (1997) also recorded similar findings. Patra*et al.*, 2008 and Deshmukh*et al.*, 2006 too reported prevalence of sickle cell patient's maximum in elderly groups and further increases with increasing age. However according to reports of Kar*et al.*, (1986) and Sahu*et al.*, (2003) reported highest frequency of sickle cell disorder in age group of 5-9 years (60.64%), While Kaur *et al.*, 1997 observed no suspects with sickle cell disease (Hb-SS) were more than 39 years. Ramasamy*et al.*, 1994 found prevalence of Hb-AS carrier was higher (47-49%) in the 10-19 year age group amongst Paniyas and Mullukurumbas lining in the western part of Nilgiris. Kar and Devi, 1997 noted presence of maximum cases in age group of 2-6 years. The maximum number detected in above 20 years might be due to growing age during which the person become self cautious and seek medical attention and treatment and thus got the diagnosis for the trait (Mohanty and Mukherjee 2002).

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Sex wise distribution of screened patients, in the studied district, shown that out of total, number of female patients was higher i.e., 4852 (62.50%) and that of males was lower i.e., 2911 (37.49%). with ratio of male to female was 728:1213 of combined Hb-SS and Hb-AS patients. In patients with genotype Hb-AS male to female ratio was 72:125 and in Hb-SS patients male to female ratio was 151:200. In a similar finding of gender-wise distribution of sickle cell anemia patients, Kar*et al.,* (1986) reported ,number of males was higher than their counter part females, and they found that there were 84 males and 47 female affected with the trait. Sahu*et al.,* (2003) found 158 (17.29%) male children and 124 (15.7%) female children were sickling positive. Brig *et al.,* (2008) observed 1336 (53%) male and 122 (47%) females. Patra*et al.,* (2008) found Hb-AS carrier pattern in 187 males (11.37%) and 20 (8.6%) in females. Deshmukh*et al., (*2006) reported prevalence of disease in males as 2.8% and that of 3.0% in females. In accordance with that of Winstrobe, 2009, in present study thesickle cell trait is more common in females than in males. The possible cause would be that the females are getting medical guidance in the backward districts of Maharashtra and treatments are medical checkups are regularly sought at the local available medical centers.

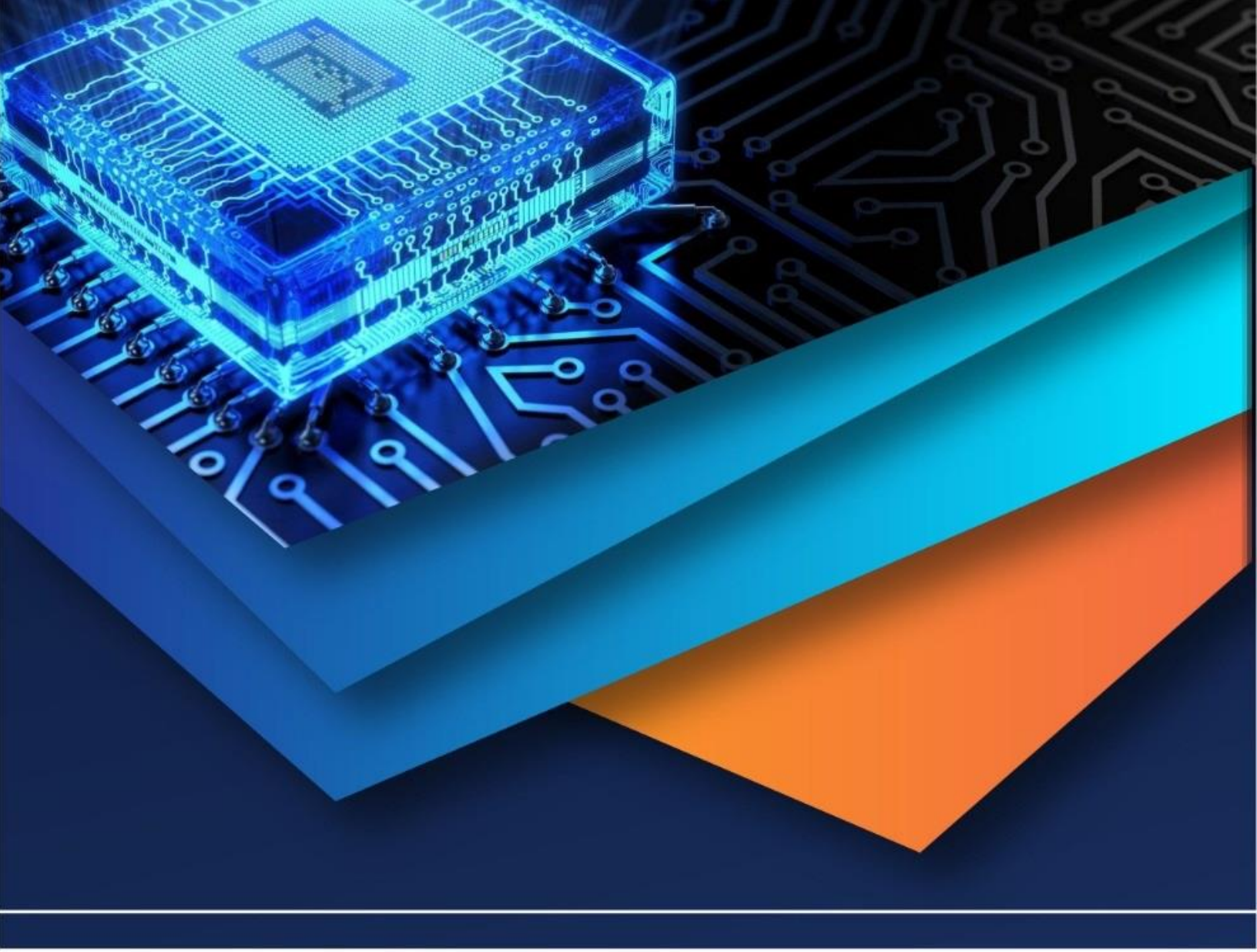
Sickle cell gene was first detected by Lehman and Cutbush (1952) among the scheduled tribes of Nilgiri Hills. Since then, more than 300 groups of scheduled tribes have been screened to look for the presence of sickle trait (Balgir 1996: Mohanty and Mukherjee 2002). The prevalence varies considerably among different tribal groups ranging from 0-35%. In certain states like Madhya Pradesh, Orissa, Chhattisgarh, Jharkhand, Gujarat. Bhatia and Rao (1987) claimed that in India, sickle cell gene is mainly restricted to tribal and scheduled caste population and found out that the carrier frequencies range is between 5-40% or more with three focal points. In accordance with this in present study total carriers Hb-AS in schedule caste 2266 (34.13%) and 488 (43.37%) were Hb-SS sufferer, among Scheduled tribe 2418 (36.43%) were carrier Hb-AS and 436 (38.75%) were sufferer Hb-SS. In other population 1954 (29.43%) were carrier Hb-AS and 201(17.81%) were sufferer Hb-SS. Rao and Goud, (1979) reported frequency of sickle cell trait ranged from 2.6 to 34.71 in tribal population. Kate and Lingojwar (2002) found over all prevalence of 10% amongst SC, ST and OBC. Sahu*et al.,* (2003), found sickling positive children were mostly belonging to scheduled caste and scheduled tribes. Deshmukh*et al.,* (2006) found high prevalence in scheduled tribes and scheduled castes as compared to other communities where it is low and rare in so called higher castes. These studies support the view that the patients of this disease is confined to economically and socially backward communities rather than other communities. The referred literature though suggested invariable castes wise prevalence but we reported that the sickle cell gene frequency is high in schedule tribe and low in other communities, of the district like Kunbi, Teli, Bhoi etc. Higher Prevalence of sickle cell gene frequency in schedule tribe may be due to practice of marriages within the community which propagates from heterozygous Hb-AS to homozygous Hb-SS state. Racial exclusion, limited opportunities and poverty are common life experiences for many such families. High fertility rates, teenage pregnancy and single parent households often reflect these neglected castes in the district. Occurrence of the trait in other communities suggested that though it is dominant 'tribal gene' it has been appeared in certain general caste groups also, may be because such population groups of in the similar district remained untouched as far as screening for sickle gene is concerned. This suggested that Sickle Cell patients are not only among the scheduled castes and scheduled tribes in India but also be suspected in subjects belonging to any caste or religion especially in the central belt of India that also included the studied district of Gadchiroli.

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