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Prevalence of sickle cell anemia Among the school students of Nagpur city

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ABSTRACT

The present paper is based on the data collected from four schools of Nagpur city under. Students from class VIIIth to Class Xth have been selected for the study. Students who were present during the time of field work were screened and venous blood from the sickle cell positive students was taken to the laboratory for confirmation test through paper electrophoresis and Capillary 2 electrophoresis. A total number of 1077 students were screened and out of which 53 were tested as carrier of sickle cell trait. Vulnerable caste groups as identified are Mahar, Teli and Kunbi. No homozygous individual has been identified in the present study.*

*The data were collected during 2010–2011, under the aegis of Anthropological survey of India project "COMMUNITY GENETIC EXTENSION PROGRAMME RELATED TO SICKLE CELL ANAEMIA AND THALASSAEMIA IN NAGPUR CITY, MAHARASHTRA'

Sickle Cell anemia is an autosomal recessive hereditary disorder wherein polymerization of hemoglobin under low oxygen tension causes structural change in the RBC. The abnormal hemoglobin----- HbS is the result of a single point mutation in the HBB gene which is responsible for hemoglobin synthesis. Because of its selective advantage against malaria, the mutant gene is largely prevalent in malaria infested regions of the world.

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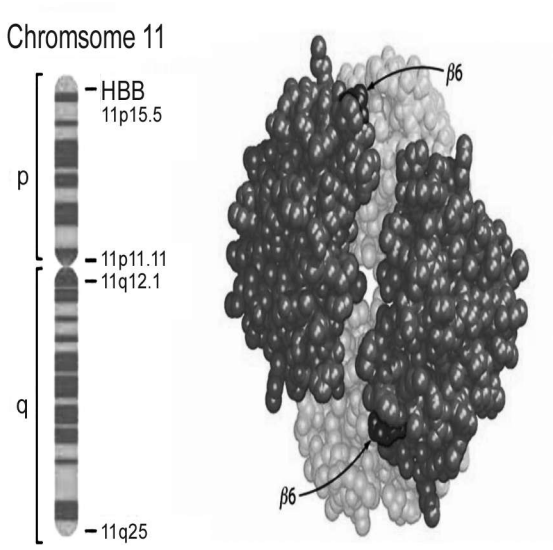

Fig. No.1: Location of HBB gene and the position of mutation in betaglobin chain of the hemoglobin

The HBB gene is located on the short arm of chromosome No.11 at 15.5 position, more precisely from 5,203,271 – 5204876 bp. Normal hemoglobin (A) consists of 2· and 2, globin chains. The mutation changes a single protein building block in beta hemoglobin, replacing glutamic acid (water soluble) to valine (fat soluble) at position 6 of the beta globin chain.(Fih-1).

This replacement produces abnormal hemoglobins which stick together and form long rigid molecules inside the RBC under low oxygen tension. These polymerized hemoglobin stick to the membrane of RBC and extends pressure and thus bend the red cell into a sickle shape.(Fig-2).

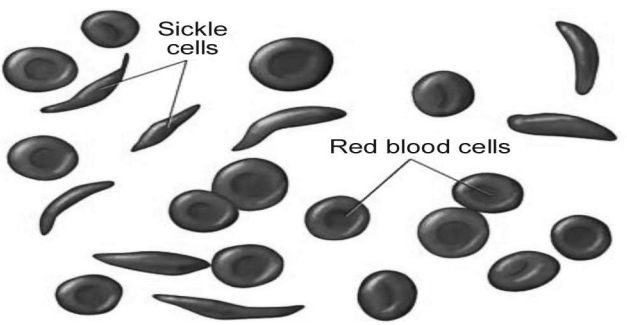

Fig. No.2: Red blood cells in heterozygous condition wherein both normal and sickle shaped RBC are visible

The Sickled Cells die prematurely and lead to the shortage of RBC and thus causes anemia. The sickled RBC loses its elasticity and can not pass through the small blood vessels causing pain and organ damage of the individual.

History

Sickle cell anemia was unknown till 1904 when an intern E. E. Iron of Chicago Presbyterion hospital found "peculiar elongated and sickle shaped cells" in the blood of W. C. Noel – a 20 years old first year dental student who was suffering from anemia. He informed Professor James Herrick – the professor of medicine of the same hospital and who brought the structural peculiarity of RBC under lime light. After nearly two decades in 1922 Verman Rason named the disease as sickle cell anemia. Linuas Pauling and colleagues were the first in 1949 to demonstrate that sickle cell disease occurs as a result of an abnormality in the hemoglobin molecule. This was the first time a disease was linked to the mutation of a specific protein – a milestone in the history in the molecular biology.

India

Sickle cell disorder was first detected in India by Lehman and Cutbush more than half a century back in 1952 among the Veddoid of Nilgiri Hills and almost at the same time by Dunlop and Majumdar among the tea garden labourers of Assam. Since then studies from different parts of India show the prevalence of the mutant gene in the whole Central India, parts of Orissa, Andhra Pradesh, Gujarat and Rajasthan.

Central India

Central India is considered as hub of this abnormal gene HbS and the distribution of the trait is 2%-35%among the population with different ethnic background. Nagpur is situated in Central India and comes under the Vidarbha region of Maharashtra state. Earliest research work on sickle cell anemia was conducted in Nagpur City and the event has silently completed its Golden Jubilee in the year 2008. In 1958 two doctors from Nagpur Medical College conducted survey among the labourers of Model Mills in Nagpur and examined them for the diagnosis of sickle cell anemia. Their results show highest incidence of SCA among the Mahar (22.2 percent) followed by Teli (11.4 percent) and Kunbi (9.4 percent) (Shukla and Solanki, 1958. Recent work among the population shows an estimated population of 400000 carriers and 22000 homozygous patients in Nagpur and other parts of Vidarbha region .This however is estimated from the annual birth rate of 260 homozygous and 7500 carriers in the Nagpur city and adjoining areas. (Srikhande 2012).

Inheritance

It was the discovery by Emmel in 1917 in the member of a family which first suggested hereditary basis of sickle cell anemia. Later on, in 1923 Huck and Sydenstricker analyzed the pedigree of the patients with sickle cell anemia and concluded that Sickle Cell Anemia

is inherited following Mendelian inheritance of recessive trait. Discovery by Pauling in 1949 regarding abnormal slow rate of migration of sickle hemoglobin on electrophoresis was confirmed in the same year by Beet through his work among the Bantu of Africa also shows that (Beet, 1949) there are two groups of sickle cell patients one is heterozygous state for sickling positive without significant symptoms and the other is symptomatic homozygous state for sickling positive with severe anemia, vaso occlusive disordes etc.

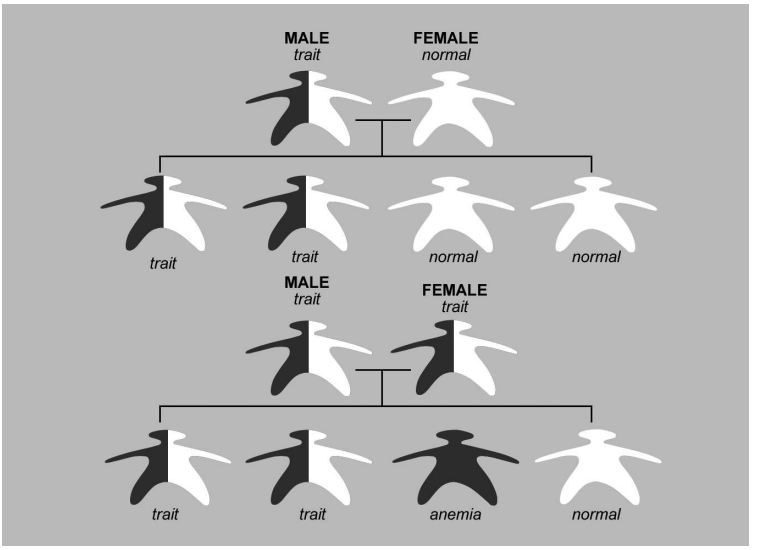

Fig No. 3: Schematic representation of inheritance of Sickle Cell anemia

Inheritance of sickle cell anemia follows the rule of Mendelian inheritance. Marriage of a normal man with a woman who is a carrier for sickle cell trait will give rise 50% normal and 50% carrier among their children; however all of them will be apparently healthy without any phenotypical abnormality. Homozygous state for sickle cell anemia arise only when two carriers (heterozygous for the trait) marries and 25% of the children becomes homozygous for the trait and suffer from severe anemia and other health problems related to sickle cell anemia. Following Mendelian inheritance 25% of them remain normal whereas 50% of them becomes carrier for the trait(Fig-3)

Material and Method

The present paper is based on the data collected from four schools of Nagpur City under the Project “. Students from Class VIII to Class X have been selected for the study as they represent adolescent premarital and age groups. Students who were present during the time of field work were screened and venous blood from the sickle cell positive students was taken to the laboratory for confirmation test through paper electrophoreses and Capillary II electrophoresis (Sebia). DNA from the blood samples of positive individuals was extracted and preserved for further study.

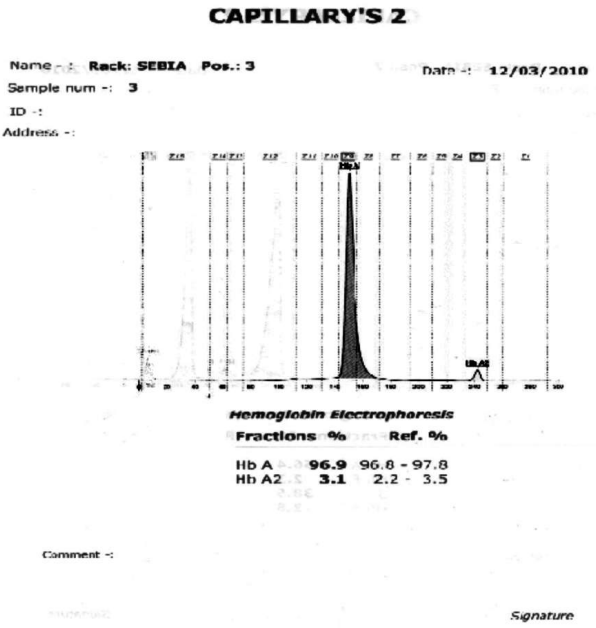

Fig. No.4 : Polygraph depicting the blood sample normal individual (AA)

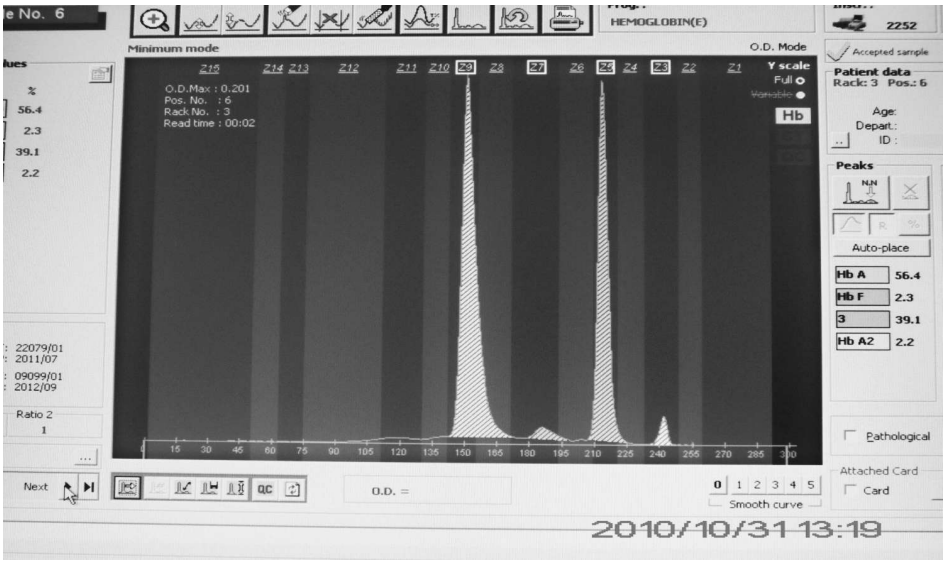

Fig. No.5: Polygraph of a heterozygous (AS) individual

Polygraph of capillary 2 electrophoresis shows the result of an individual who was tested as screened positive but actually confirmed as false positive after the analysis of blood sample in the capillary 2 electrophoresis. (Fig.. 4). Polygraph of a heterogygous sample (Fig--5) clearly shows the graph in Zone 5 region which is the position for hemoglobin S.

Awareness program have been conducted among the students of four schools with the help of audio visual presentation. Impact of the lecture has also been assessed by questioning the students regarding their perception about inheritance, social stigma etc. of sickle cell disease immediately after the presentation. Booklets on sickle cell have also been distributed among the interested students

Results and Discussion

A total number of 1077 students were screened, out of which nearly 5 % percent were tested with confirmation as sickle cell positive. No homozygous individual has been identified in the present study. Out of total – 64 communities, majority of the students belong to OBC category followed by Scheduled Caste, General, Scheduled Tribe and Nomadic Tribal Groups (Fig-6).

Table No.1
Population Composition of school students under present study

Sr. No	Caste	Category	Male	Female	Total
1	Mali	OBC	28	12	40
2	Sutar	OBC	4	21	25
3	Gondhali	OBC	2	0	2
4	Lohar	OBC	16	7	23
5	Wadhi	OBC	7	9	16
6	Kewat	OBC	0	2	2
7	Jain	Gen	2	3	5
8	Chambhar	SC	5	7	12
9	Mochi	SC	0	3	3
10	Bahana	SC	1	0	1
11	Padam Sali	OBC	1	0	1
12	Kori	SC	1	0	1
13	Khargarbhat	SBC	15	9	24
14	Koshti	SBC	15	9	24
15	Balai	SC	4	2	6
16	Dhobi	Gen	7	6	13
17	Barai	OBC	3	7	10
18	Bhat	OBC	2	7	9

Sr. No	Caste	Category	Male	Female	Total
19	Mang	SC	5	3	8
20	Matang	SC	1	0	1
21	Burud	SC	0	1	1
22	Mehter	SC	1	0	1
23	Banjari	VJ	4	0	4
24	Pinjari	OBC	0	1	1
25	Dalal	Gen	0	1	1
26	Brahman	Gen	4	9	13
27	Christian	Gen	0	1	1
28	Rajput	Gen	1	2	3
29	Thakur	ST	4	5	9
30	Dhangar	NT	1	1	2
31	Beldar	NT	1	0	1
32	Gadi Lohar	NT	14	8	22
33	Godhal	NT	0	4	4
34	Gaoli	Gen	0	1	1
35	Rangari	OBC	0	1	1
36	Dhiwar	NT	6	3	9
37	Maratha	Gen	10	10	20
38	Muslim	Gen	12	4	16
39	Teli	OBC	89	80	169
40	Pawar	OBC	13	18	31
41	Kunbi	OBC	86	94	180
42	Halba	ST	9	18	27
43	Sonar	OBC	21	20	41
44	Bhoi	NT	3	2	5
45	Kumbhar	OBC	0	1	1
46	Navi	OBC	4	9	13
47	Gond	ST	13	13	26
48	Shimpi	OBC	6	9	15
49	Mahar	SC	79	77	156
50	Ahir	NT	3	2	5

Sr. No	Caste	Category	Male	Female	Total
51	Kalar	OBC	22	16	38
52	Wadhai	Gen	1	1	2
53	Kushwa	Gen	2	2	4
54	CKP	Gen	2	2	4
55	Halwai	Gen	1	1	2
56	Nathjogi	SC	1	1	2
57	Gowari	OBC	3	2	4
58	Khatik	OBC	2	1	3
59	Wadi	OBC	1	1	2
60	Mana/Mane	ST	1	1	2
61	Gurav	OBC	0	3	3
62	Raut	OBC	1	0	1
63	Satnami	SC	1	0	1
64	Peraki	OBC	2	1	3
		TOTAL	**543**	**534**	**1077**

Community wise distribution of the data shows that the students of the four schools are distributed in 64 communities. The majority of which are Teli, Kunbi and Mahar.

Table no.2

Distribution of total population in different categories under present study

Sr. No.	Category	Male	Female	Total	%
01.	OBC	313	321	634	58.87
02.	SC	99	94	193	17.92
03.	ST	27	37	64	5.94
04.	Gen	42	44	86	7.98
05.	VJ	4	0	4	0.37
06.	NT	28	20	48	4.46
07.	SBC	30	18	48	4.46
	TOTAL	**543**	**534**	**1077**	
	Percentage	**50.42%**	**49.58%**		**100%**

Table No.2 depicts highest number of OBC (58.8 percent) and lowest number of nomadic tribe (0.37 percent). Next to OBC major category of students under the presents study is represented by Scheduled Caste (17.92 percent), General Caste group (7.98 percent) and Scheduled Tribe (5.94 percent). Nomadic Tribe and Special Backward Class shares equal representation (Fig-6)) under the current study.

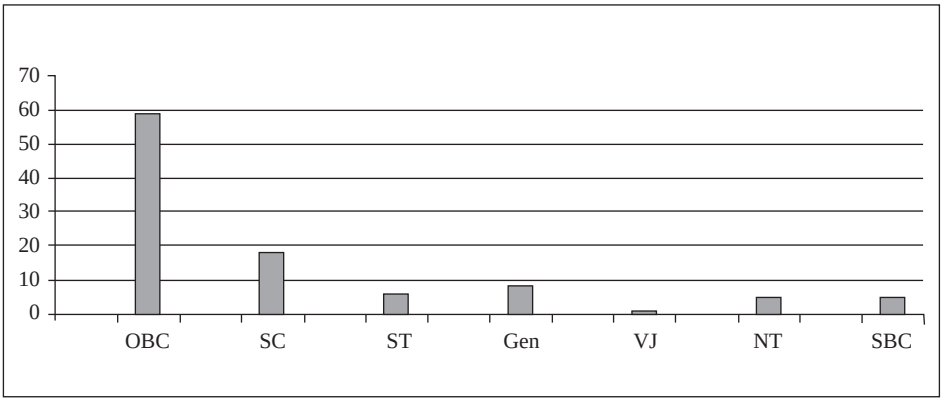

Fig. No.6: Distribution of students under different categories

Incidences of sickle cell trait (AS) among the OBC and SC are equal and show higher percentage (2.13 percent) as compared with the students under ST, SBC and General category. (Fig. No.7).

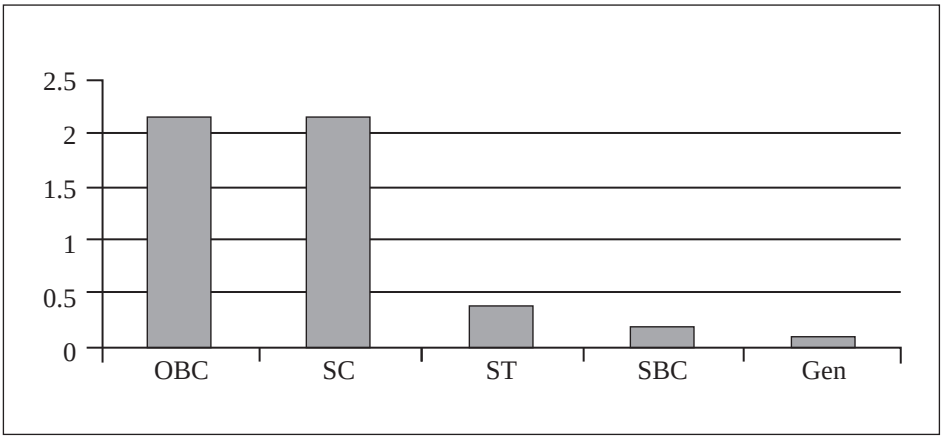

Fig. No.7: Distribution of sickle cell trait among the students under different categories

Table No.3

Distribution of sickle cell carries (as) in different caste groups

S.No	Caste	Category	Male	Female	Total	%
01	Lohar	OBC	0	1	1	0.09
02	Mahar	SC	6	17	23	2.13
03	Kunbi	OBC	2	3	5	0.46
04	Barai	OBC	1	0	1	0.09
05	Teli	OBC	5	4	9	0.83
06	Gowari	SBC	2	0	2	0.18
07	Kalar	OBC	2	0	2	0.18
08	Sonar	OBC	1	0	1	0.09
09	Gond	ST	2	1	3	0.28
10	Dhiwar	NT	0	1	1	0.09
11	Sutar	OBC	0	1	1	0.09
12	Halba	ST	0	1	1	0.09
13	Power	OBC	0	1	1	0.09
14	Mali	OBC	0	1	1	0.09
15	Wadai	OBC	1	0	1	0.09
		TOTAL	**22**	**31**	**53**	

Community wise distribution of data reveal highest incidence of sickle cell trait among the Mahar (Fig-8) as compared to other communities.

Distribution of sickle cell trait among the other communities show more or less same pattern i.e. less than 0.5 percent excepting Teli (0.83 percent).(Table No.3).

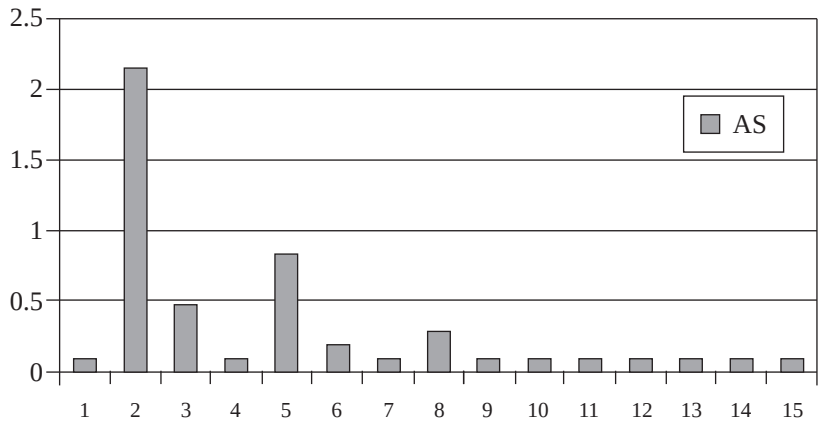

Fig. No.8: Distribution of sickle cell trait among the major cast groups under study (Graph corresponding to Table No.3)

Table no.4

Percentage showing intra cast incidences of sickle cell trait

Sr. No.	Caste	Total Sample (M + F)	AS Individual	Percentage
1	Mali	40	1	2.5
2	Sutar	25	1	4
3	Gondhali	2	0	0
4	Lohar	23	0	0
5	Wadhi	16	1	6.25
6	Kewat	2	0	0
7	Jain	5	0	0
8	Chambhar	12	0	0
9	Mochi	3	0	0
10	Bahana	1	0	0
11	Padam Sali	1	0	0
12	Kori	1	0	0
13	Khargarbhat	24	0	0
14	Koshti	24	0	0
15	Balai	6	0	0
16	Dhobi	13	0	0
17	Barai	10	1	10
18	Bhat	9	0	0
19	Mang	8	0	0
20	Matang	1	0	0
21	Burud	1	0	0
22	Mehter	1	0	0
23	Banjari	4	0	0
24	Pinjari	1	0	0
25	Dalal	1	0	0
26	Brahman	13	0	0
27	Christain	1	0	0
28	Rajput	3	0	0
29	Thakur	9	0	0
30	Dhangar	2	0	0
31	Beldar	1	0	0
32	Gadi Lohar	22	0	0
33	Godhal	4	0	0
34	Gaoli	1	0	0

Sr. No.	Caste	Total Sample (M + F)	AS Individual	Percentage
35	Rangari	1	0	0
36	Dhiwar	9	1	11.11
37	Maratha	20	0	0
38	Muslim	16	0	0
39	Teli*	169	9	5.32
40	Pawar	31	1	3.22
41	Kunbi*	180	5	2.76
42	Halba	27	1	3.7
43	Sonar	41	1	2.44
44	Bhoi	5	1	20
45	Kumbhar	1	0	0
46	Navi	13	0	0
47	Gond	26	3	11.54
48	Shimpi	15	0	0
49	Mahar*	156	23	14.74
50	Ahir	5	0	0
51	Kalar	38	2	5.26
52	Wadhai	2	1	50
53	Kushwa	4	0	0
54	CKP	4	0	0
55	Halwai	2	0	0
56	Nathjogi	2	0	0
57	Gowari	5	2	40
58	Khatik	3	0	0
59	Wadi	2	1	50
60	Mana/Mane	2	0	0
61	Godavi	3	0	0
62	Raut	1	0	0
63	Satnami	1	0	0
64	Peraki	3	0	0

Intra community comparison of data of those communities which have significant representation (more than 100 individuals) in the total number of collected data shows highest incidence of sickle cell trait among the Mahar (14.74 percent) followed by Teli (5.32 percent) and Kunbi (2.74 percent). This however supports the results of the first study in Nagpur by Shukla and Solanki (1958) and reiterates the categorical position of the aforesaid three caste groups with regards to sickle cell trait. Research works from

different parts of Maharashtra in different time period also depict the same view (Table No. 5). It is important to mention that the percentage of sickle cell trait has declined in all these three communities under the current study. Higher percentage of sickle cell trait among some other communities has not been taken in to consideration because of their small number of representation in the present study.

Table no. 5

Prevalence of sca in a few population of maharashtra

Sr. No.	Community	Incidence (%)	Reference & Year
01.	Bhil	15.85	Negi, 1976
02.	Bhil	18.00	Negi, 1978
03.	Bhil	20.24	Sathe, etal, 1987
04.	Bhil	20.60	Kate, 2001
05.	Pradhan	09.00	Ahmed & Choudhary, 1980
06.	Pradhan	11.08	Bankar etal, 1984
07.	Pradhan	10.60	Deshmukh etal, 2006
08.	Pradhan	15.80	IGIMC, Wardha, 2009
09.	Teli	11.10	Shukla & Solanki, 1958
10.	Halba	13.6	Negi, 1976
11.	Gond (MP)	19.4	Negi, 1963
12.	Mahar (Rural)	18.6	Urade, et. al, 2001
13.	Mahar (Nagpur)	18.1	Das et. al, 1961
14.	Pardhan (Nanded)	16.8	Banker, et. al, 1984
15.	Pawar	25.5	Kate, 2001
16.	Halba (Raipur)	13.6	Negi, 1976
17.	Thakur	6.06	Tiwari, 1980
18.	Chamar (Raipur)	4.5	Tiwari, 1980
19.	Chamar (Raipur)	6.7	Tiwari, 1980
20.	Mahar (Raipur)	18.8	Tiwari, 1980
21.	Mahar (Raipur)	19.5	Tiwari, 1980
22.	Gond (Raipur)	7.96	ICMR, 1986
23.	Kamar (Raipur)	2.38	Tiwari, 1986

Conclusion

Community specific trend of occurrence of sickle cell trait in Nagpur in particular and Maharashtra as a whole is discussed in the present study. The percentage of incidences however shows trend of decline among the vulnerable caste groups (Mahar, Teli and Kunbi) which may be a sign growing awareness among these groups regarding the particular genetic trait.

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