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Hemoglobinopathy pattern among school and college students in Marathwada region of Maharashtra: A study of 500 students

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Abstract
Introduction: Sickle hemoglobin is highly prevalent among the tribal of central, southern, and western India which ranges from 10 to 23%. Sickle cell hemoglobinopathy a hereditary conditions that causes morbidity and mortality in school going children.
Aims and Objectives: To study the prevalence of sickle cell disorder among the students.
Material and Methods: All the patient presented with sign and symptoms of anemia with suspicious of sickle cell disease and visiting various schools and colleges the samples were collected for CBC, peripheral smear, HPLC examination and reported.
Results: Total 500 blood samples collected from school and college students for screening by HPLC and their hemoglobinopathy status. Showing prevalence of sickle cell anemia is 5.4%.
Conclusions: It can be concluded that approaching community can be possible through school going adolescent for conducting surveillance of sickle cell anaemia. As it was small study, similar study of large size can be repeated by regular and planned visit to schools.

Keywords: Sickle, students, hemoglobinopathy

Introduction
Sickle cell disease is an autosomal recessive genetically transmitted hemoglobinopathy responsible for considerable mortality and morbidity. It is prevalent in many parts of the India, where the prevalence in different communities has ranged from 9.4-22.2% [3]. It is one of the most common hereditary disease occurring worldwide, which may affects any organ or system of the body. It is irreversible, untreated health problem predominantly seen amongst various tribes [10].

India caters to nearly 20 million peoples with Sickle cell disease. The Sickle cell gene in India was first described among tribal groups in the south India, but the recent data unfolds that the disease is not restricted only to tribal belts but is widely prevalent and has penetrated different casts and communities in our country. The average frequency of Sickle cell disease in India is 4% [7].

The overall prevalence of Sickle cell disease in different tribal population in the state of Maharashtra is 10% for carrier state and 0.5% for the sufferers. The prevalence is very high amongst the tribal population groups from Nandurbar and Gadchiroli district of the state [13].

The schools are sacred because they provide an environment for learning skills and for development of intelligence that can be utilized by students to achieve their goals in life. “To learn effectively, children need good health’ so, health is key factor for school entry, as well as continued participation and attainment in school. If we fail to nurture the health of school children there can be immense destabilizing effects on political, social and economic system. In Maharashtra residential schools are mainly for the students belongs to schedule tribes and these are located in tribal areas of the state [15].

The present study was carried out to screen school going children and adolescent students, which is the effective medium to reach family and community for the screening for Sickle cell disease.

Aims and objectives
To study the prevalence of sickle cell disorder among the students, and to correlate clinical

symptomatology with hematological parameters.

Material and Methods

This is cross sectional analytical study. After the ethical committee approval, the patients presented with sign and symptoms of anemia with suspicious of sickle cell disease that had attended pediatric/medicine OPD and visiting various schools and colleges which fulfilling inclusion and exclusion criteria the samples were collected. The present study was carried out in the Department of Pathology at our institute.

Inclusion criteria

Students irrespective of their origin, caste and ethnic background having sign and symptoms of anemia were included.

Exclusion criteria

1. Students who had received blood transfusion within three months prior to the day of inclusion.
2. Students above the age of 30 years.
3. Students with co-morbidity such as pre-existing chronic illness other than sickle cell disease were excluded from the study.

Study designs

All samples collected were tested for CBC, peripheral smear, HPLC according to standard protocol with Automated High Performance Liquid Chromatography (HPLC) Bio-Rad Sickle Cell Anemia Short Programme. The Sysmex XS-1000i is an automatic multi-parameter cell counter for CBC.

Statistical analysis

Statistical analysis was done with the help of Microsoft excel, Office 2003 version software.

Results

The present study was carried out from October 2010 to November 2012. Total 500 blood samples collected from school and college students for screening by HPLC for their hemoglobinopathy status. The lowest age was 5 years and eldest was of 30 years and following observations were made:

Table 1: Showing prevalence of sickle cell anemia

Total study subjects	Total cases of SCD	Percentage
500	27	5.4%

The prevalence of sickle cell disease among students was 5.4%.

Total 500 cases of blood samples were tested, out of which 354 cases (70.8%) having normal hemoglobin pattern and 146 (29.2%) cases were having abnormal hemoglobin pattern

Table 2: Sex-wise distribution of total cases (n=500)

Hb pattern	Male		Female		Total
	Cases	Percentage	Cases	Percentage	
Normal	160	64.2	194	77.2	354
Abnormal	89	35.7	57	22.7	146
Total	249	99.9	251	99.9	500

Table 2 shows sex wise distribution of 500 cases, out of 146 cases with abnormal hemoglobin pattern, males were 89 (35.7%) and females were 57 (22.7%)

Table 3: Age wise distribution of abnormal pattern in hemoglobinopathies (n=146)

Abnormal Hb Patterns	Age (Yrs.)				
	5-10	11-15	16-20	21-25	26-30
SCD	12	7	5	2	1
SC Trait	2	7	7	12	7
SC Trait with ß. Thal Trait	1	1	2	-	2
ß. Thal. major	2	-	-	1	-
ß. Thal Trait	6	4	16	23	14
SCD with ß. Thal Trait	4	3	3	2	-
Total	27	22	33	40	24
Percentage	18.5%	15%	22.7%	27.4%	16.4%

Table 3 shows, total 146 cases of abnormal hemoglobinopathies studied, of which 40 cases (27.4%) were observed in age group 21 to 25 years, followed by 33 cases (22.7%) in age group between 16 to 20 years.

Abnormal hemoglobin pattern wise distribution of 146 cases, of which 63 cases (43.15%) were ß. Thal Trait followed by 35 (24%) cases were sickle cell trait, 27 (18.49%) sickle cell disease, 12 (8.21%) were sickle cell disease with ß. Thal Trait, 6 cases (4.1%) were sickle cell trait with ß. Thal Trait and ß. Thal. Major were observed in 3 (2.05%) cases

Table 4: Caste wise distribution of abnormal hemoglobinopathies cases (n=146)

Caste	No. of cases	Percentage
Schedule caste	57	39.0
Schedule tribe	09	06.1
Nomadic tribe	18	12.3
Other backward	13	09.0
Open category	46	31.5
Other	03	02.1
Total	**146**	**100.0**

Table 4 shows maximum cases were found in schedule caste community i.e. 57 cases (39%), followed by 46 cases (31.5%) in open category

Table 5: Ethnic Group wise and pattern wise distribution of abnormal hemoglobinopathy cases (n=146)

Caste	Ethnic group	SCD	SC Trait	SCD with ß. Thal. trait	SC trait with ß. Thal. trait	ß. Thal. Major	ß. Thal. trait	Total
SC (n=57)	Bauddha	5	14	3	-	1	14	37
	Chambhar	-	1	-	-	-	-	1
	Mahar	3	2	-	2	-	4	11
	Mang	1	1	-	-	-	2	4
	Nandiwale	1						1
	Nhavi	-	-	1			1	2

	Walmik	-	-	-	1			1
ST (n=9)	Bhil						1	1
	M. Koli	3	-	-	1	-	3	7
	Munnerwar Lu						1	1
NT (n=18)	Banjara	2	3	4	-	-	3	12
	Dhangar	1	1					2
	Lamani						1	1
	Rajput						1	1
	Vanjari		1				1	2
OBC (n=13)	Gaowli	-	1					1
	Hatkar						1	1
	Mali						2	2
	Padmashali						1	1
	Sutar	1	2				1	4
	Telgu	1	1					2
	Teli						2	2
Open (n=46)	Brahmin						6	6
	Jain						1	1
	Maratha	1	2	-	-	1	4	8
	Muslim	8	5	4	-	1	13	31
Other (n=3)	Bandra	-	-	1			1	2
	Upadhaya	-	1					1
Total		27	35	13	4	3	64	146

Table 5 shows, ethnic group wise and pattern wise distribution in abnormal hemoglobinopathies of 146 cases studied. In Schedule caste category total 57 cases seen. Out of which Bauddha community had 37 cases among which Sicle Cell trait and ß thal trait i.e. 14 cases each and 11 cases in Mahar community in which ß Thal trait were observed in 4 cases, Sickle Cell Disease in 3 cases, SC trait and SC trait with ß Thal trait in 2 cases each.

Amongst open category (n=46), 31 cases belongs to Muslim category having ß Thal trait 13 cases followed by, 8 cases of Sickle Cell Disease, 5 cases of SC trait, 4 cases of SCD with ß Thal trait and only one in ß Thal major.

In Nomatic tribes of 18 cases, maximum cases were observed in Banjara community i.e. 12 cases. Out of which SCD with ß Thal trait seen in 4 cases, followed by SC trait and ß Thal trait in 3 cases each and SCD in 2 cases.

In other backward class 13 cases were observed, of which maximum cases observed are ß Thal trait i.e. 4 cases in Sutar community.

Amongst Schedule tribe caste total 9 cases were observed. Out of which 7 cases belongs to M. Koli community, of which SCD and ß Thal trait were observed in 3 cases each. & 1 case of SC trait witb ß thal trait. While 2 cases of ß thal trait seen in remaining category.

Hematological profile of Sickle Cell Trait with ß Thal trait

HPLC parameters like HbF, HbA0, HbA2 and Hbs in 6 cases (4.1%) of Sickle Cell Trait with ß Thal trait cases studied. HbS level and HbA2 level were increased.

Hematological profile of Sickle Cell Disease with ß Thal trait (n=12)

HPLC parameters like HbF, HbA0, HbA2 and Hbs in 12 cases (8.21%) of Sickle Cell Disease with ß Thal trait cases studied. HbF and HbS both were raised.

Hematological profile of Sickle Cell Disease (n=27)

HPLC parameters like HbF, HbA0, HbA2 and Hbs in 27 cases (18.49%) of Sickle Cell Disease cases studied. HbF and HbS both levels were increased.

Hematological profile of Sickle Cell Trait (n=35)

HPLC parameters like HbF, HbA0, HbA2 and Hbs in 35 cases (24%) Sickle Cell trait cases studied. HbA were increased and HbS were below 50.

Hematological profile of ß Thal. Major (n=3)

HPLC parameters like HbF, HbA0, HbA2 and Hbs in 3 cases (2.05%) of ß Thal. Major cases studied. HbF raised upto 90 and HbA2 will be normal or raised.

Hematological profile of ß Thal. Trait (n=63)

HPLC parameters like HbF, HbA0, HbA2 and Hbs in 63 cases (43.1%) of ß Thal. trait cases studied. HbF were increased upto 5% and HbA2 were raised up to 8

Discussion

Present study was a prospective study of 500 school and college going students between age group 5 to 30 years, with prevalence of sickle cell disease was 5.4%.

Table 6: Comparative study of prevalence of sickle cell disease

Author (Year)	Prevalence (%)
Kamble and Chatruvadi (1999) [3]	5.7%
Bipin Vasava (2009) [10]	25.5%
Patel *et al.* (2011) [14]	11.37%
Dhumne *et al.* (2011) [12]	18.3%
Gunjal *et al.* (2012) [15]	6.28%
Present study (2012)	5.4%

Table 6 shows our findings are correlate with the prevalence showed by Kamble and Chatruvedi (1999) [3] and Gunjal *et al.* (2012) [15].

The distribution of hemoglobin pattern concern, in the present study 354 (70.8%) cases were normal and 146 (29.2%) were abnormal pattern. Gunjal S. (2012) [15] showed maximum cases i.e. 850 (93.6%) normal hemoglobin pattern and 58 cases (6.3%) with abnormal pattern in their study of 908 cases at Residential Tribal School, Central India. Our

findings were more as compared to incidence showed by Gunjal S. (2012) [15]

Table 7: Comparative study of male to female ratio with previous studies

Author (Year)	Male	Female	Ratio
Sahu *et al.* (2003) [8] 6 (n=1704)	914 (53.6%)	790 (46.3%)	1.15:1
Gunjal *et al.* (2012) [15] (n=908)	463(50.9%)	445 (49.1%)	1.03:1
Patel *et al.* (2012) [14] (n=168495)	85862(50.9%)	82633(49.1%)	1.03:1
Present study (2012) (n=146)	89 (61%)	67 (39%)	1.56:1

Thus in all the previous studies the ratio of male to female were more or less similar. Our results were well correlated with Gunjal S. (2012) [57] and Patel A. (2012) [14] studies, who showed same male to female ratio.

In the present study, out of 146 abnormal cases, male cases were predominant over female i.e. 89 (61%) male and 57 (39%) were female with male to female ratio 1.56:1.

Table 8: Comparative study of age wise distribution of abnormal cases with previous studies

Author (Year)	Age group				
	5-10	11-15	16-20	21-25	26-30
Sahu *et al.* (2003) [6] (n=1704)	171	22	-	-	-
Dumne *et al.* (2012) [12] (n=7)		2		5	
HIHF/GAH Sickle cell disease center Nilgiri District South India [19] (n=34)	17	13		4	
Present study (2012) (n=146)	27	22	33	40	24

Sahu T. (2003) [6] showed, out of 1013 cases between age group 5 to 15 years, of which maximum cases i.e. 171 (16.8%) were between 5 to 9 years and 22 (2.17%) cases were between age group 10 to 15 years. Ours study correlates with the findings showed by Sahu T. (2003) [6] at Berhanpur, Orissa.

In the present study abnormal hemoglobin pattern wise distribution of 146 cases, of which 63 cases (43.15%) were ß. Thal Trait followed by 35 (24%) cases were sickle cell trait, 27 (18.49%) sickle cell disease, 12 (8.21%) were sickle cell disease with ß. Thal Trait, 6 cases (4.1%) were sickle cell trait with ß. Thal Trait and ß. Thal. Major were observed in 3 (2.05%) cases.

Brandelise *et al.* (2004) [7], in their study of major haemoglobin phenotypes among 281,884 samples during period 1992 to 2000 at Sao Paulo State, Brazil showed sickle cell trait in 5197 cases 1.98% and HbC Trait in 1615 (0.57%) cases followed by Homozygous sickle cell disease in 29 cases, sickle cell hemoglobin C disease in 26 cases and only sickle cell ß thalassemia in 2 cases.

Patel *et al.* (2012) [14] showed prevalence of ß. Thal Trait and sickle cell trait in total 1,68,495 cases of which they showed 2,935 (1.78%) cases of ß. Thal Trait and sickle cell trait in 19,154 (11.37%) cases.

Abnormal hemoglobinopathies does not correlates with the other studies as their number of cases were more.

In our study, as far as caste wise distribution of abnormal hemoglobinopathies cases, maximum cases were found in schedule caste community i.e. 57 cases (39%), followed by 46 cases (31.5%) in open, in Nomadic tribe 18 cases (12.3%), 13 cases in other backward group, 9 (6.1%) cases in schedule tribe and only 3 cases in other ethnic group.

Kate S.L. and Lingojwar D.P. (2002) [5] showed incidence of sickle cell disorder amongst scheduled tribe population group in Maharashtra state. They showed 196 (51.4%) cases in scheduled tribe category followed by 144 (37.7%) in scheduled caste and 41 cases (10.7%) in other backward class in their 381 cases studied.

Balgir R.S. (2006) [16] showed in their study of 667 cases, general caste in 64.6% cases of sickle cell disorder, followed by scheduled caste 27.4% cases of 79.6% ß thalassemia syndrome and 91.3% other hemoglobinopathies. Whereas in Scheduled caste, 27.4% sickle cell disorder followed by 16.2% ß thalassemia syndrome and 8.7% other hemoglobinopathies. In scheduled tribe community, 8% sickle cell disorder, 4.2%% ß thalassemia syndrome and no case was found in other hemoglobinopathies.

Patel *et al.* (2012) [14] showed distribution of sickle cell and ß thal trait in different caste of Gujarat. They showed scheduled caste community had 2.64% to 10.52% range, followed by scheduled tribe between range 1.12% to 18.65% and in general caste ranges from 1.03% to 15.63%. Hence, scheduled caste community had more prevalence in our study. Kate and Lingojwar (2002) [5] also showed maximum cases in scheduled caste community and our results were correlate with the finding showed by Kate and Lingojwar (2002) [5]. In our study, open category had also high prevalence. But we cannot compared our results with previous studies as no study showed open category prevalence rate. About scheduled tribe community our results were more or less correlate with the findings showed by Bulgir R.S. (2006) [16] and Patel *et al.* (2012) [16].

In the present study, ethnic group wise and pattern wise distribution in abnormal hemoglobinopathies of 146 cases studied. Schedule caste category was more i.e. 57 cases in which Bauddha community had more cases in SC trait and ß thal trait i.e. 14 cases each and 11 cases in Mahar community in which ß Thal trait were observed in 4 cases, SCD in 3 cases, SC trait and SC trait with ß Thal trait in 2 cases each. Amongst open category, out of total 31 cases, Muslim category was most common in ß Thal trait i.e. 13 followed by, 8 cases in SCD, 5 cases in SC trait, 4 in SCD with ß Thal trait and only one in ß Thal major.

In Nomatic tribes of 18 cases, maximum cases were observed in Banjara community i.e. 12 cases. SCD with ß Thal trait 4 cases, followed by SC trait and ß Thal trait 3 cases each and SCD in 2 cases. In other backward class 13 cases were observed, of which maximum cases observed in ß Thal trait i.e. 4 cases in Sutar community.

Amongst Schedule tribe community, total 9 cases were observed, whereas out of maximum 7 cases, SCD and ß Thal trait were observed in 3 cases each in M. Koli community. Only 3 cases were observed in other category. Gorakshakar A.C. (2006) [8] showed maximum prevalence of sickle cell disease in Mahar community i.e. 5.14% followed by Brahmin, Muslim and Maratha and Koshti in few cases of open category. Balgir R.S. (2006) [16] showed Teli, Rajput, Muslim and Brahmin in open category showed maximum prevalence in Teli i.e. 2.1%, followed by Rajput in 1.7%, Muslim and Brahmin in 1.3% each. Zade V.S. *et al.* (2011) [13] gives prevalence only in scheduled tribe category, which Gaoli was observed in 0.4% cases.

Patel *et al.* (2012) [14] showed Brahmin in 1.98% in S.C. trait and 3.49% in ß thal trait. Rajput 7.7% in S.C. trait and 2.9%

in ß thal trait among open category. Chambhar was the common subcaste in scheduled caste category, which shows 10.52% S.C. trait and 3.49% ß thal trait, Mahar (Harijan) 8.1% in S.C. trait and 3.5% in ß thal trait. Koli the only category that was correlated with our study was 16.87% S.C. trait, no case of ß thal. Trait were found in their study.

In the present study also Mahar community had more prevalence i.e. 7.5% which was also correlated with previous studies who showed more or less prevalence in Mahar (Harijan) community i.e. Gorakshakar A.C. (2006) [8]-5.14% and Patel *et al.* (2012)-8.1%.

In the present study, mean of hematological parameters i.e. Hb%, PCV, TLC, Platelet, MCV, MCH, MCHC and HPLC parameters like HbF, HbA0, HbA2 and Hbs in 27 cases (18.49%) in sickle cell disease cases were studied

Table 9: Comparison of mean CBC parameters in SCD cases with previous studies.

Author (Yrs.)	CBC Parameters (mean)						
	Hb%	**PCV**	**TLC**	**PLT**	**MCV**	**MCH**	**MCH C**
Mohd. Zailaie *et al.* (2003) [2]	10.32	25.26	79.51	-	33.34	-	-
Mohanty *et al.* (2008) [9]	8.7	-	-	-	72.5	23.3	32.2
Sukla R.N. *et al.* (/1958/2009) [11]	8.34	21.8	-	-	93	25	26.4
Present study (2012)	6.4	18.9	8422	2.33	70.6	28.3	32.6

In the present study, our CBC parameters in SCD cases were more or less comparable with that of Mohanty *et al.* (2008) [9] except hemoglobin level and MCH level

Conclusions

So to conclude with screening among open category is mandatory to rule out hemoglobinopathy and early screening among students is necessary for early management. Premarital screening is necessary to reduce further complications. That gives idea to family screening to reduce further burden of management & complication. School going adolescents can be an effective medium to reach the community for the screening of the people for sickle cell anaemia.

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