pISSN: 0976 3325 eISSN: 2229 6816

ORIGINAL ARTICLE

PREVALENCE OF COMMON HEMOGLOBINOPATHIES IN GUJARAT: AN ANALYSIS OF A LARGE POPULATION SCREENING PROGRAM

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ABSTRACT

Objectives: To determine prevalence and geographical distribution of ß-thalassemia trait and sickle cell trait in Gujarat

Methods: The present study includes 3, 17,539 persons who were screened for hemoglobinopathies from September 2004 to November 2008. Blood samples were collected into EDTA and run in coulter AC.T diff 2 TM, electronic analyzer and analyzed on Bio-Rad Variant I HPLC System.

Results: The prevalence of ß-thalassemia trait (BTM) and sickle cell trait (SCT) is 1.95% and 6.54% respectively. In tribal area, 1.74% has BTM and 11.37% has SCT trait while in non-tribal area, BTM is seen in 2.18% and 1.1% has SCT. Rathva (18.65%), Nayka (15.38%), Gamit (17.64%), Tadvi (16.95%), Bariya (16.5%), Adivasi (15.25%) and Chaudhary (15.63%) communities have higher prevalence of SCT. BTM is more prevalent in Bhanusali (8.1%), Bhakta (7.93%) and Luhana (6.5%) community. SCT is very common in schedule castes (8.1-10.52 %) and it is seen in many general castes.

Conclusion: The prevalence of BTM is 1.95% while SCT is 6.54% in Gujarat. Sickle cell gene frequency is high in tribals (11.37 %) except in Banaskantha (5.12 %) and Sabarkantha (3.43%). Sickle cell is prevalent in all schedule castes and many general castes. ß-thalassemia gene frequency is higher in Sindhi (Bhanusali) and Bhakta communities and in Banaskantha district (4.45%).

Keywords: Abnormal hemoglobin, Hemoglobinopathies, Prevalence, Screening, Sickle cell, Thalassemia

INTRODUCTION

Hemoglobinopathies are a group of genetic disorders of hemoglobin. They affect 4.5% of the world population.[1] The prevalence of β-thalassemia trait and sickle cell in India varies between 3-17% and 1-44% respectively because of consanguinity and caste and area endogamy.[2] Every year, ten thousand children with β-thalassemia major are born in India, which constitutes 10% of the total number in the world.[3] Inherited hemoglobin disorders are an important cause of morbidity and mortality. The curative treatment like bone marrow transplantation is costly and so prevention is the cost effective strategy, which includes population screening, genetic counseling and prenatal diagnosis.

There is no population based large study from Gujarat. Our study is the largest screening study in India and hence estimates prevalence of various hemoglobin disorders more accurately. The study is a part of thalassemia and sickle cell prevention program run by Indian Red Cross Society- Gujarat State Branch with the financial support from the Gujarat government.

MATERIAL AND METHOD

The present study of the prospective voluntary screening programme was conducted under the auspices of the Gujarat State Branch of Indian Red Cross Society, Ahmedabad, covering the period from September 2004 to November 2008. Blood samples from a total of 3,17,539 persons (1,68,495 from tribal area and 1,49,044 from non-tribal area) were collected from different areas of Gujarat which includes Banaskantha, Baroda, Bharuch, Dahod, Narmada, Panchmahal, Sabarkantha, Ahmedabad, Anand, Gandhinagar, Bhavnagar, Amreli, Jamnagar, Kheda, Kutch, Navsari, Patan, Rajkot, Surat, Porbandar, Surendranagar and Valsad districts. For variant study, collection of Blood samples were done according to practical manual of Dacie.[4] The blood samples were mixed on electric blood mixture for 8 to 10 minutes and thereafter analyzed on the coulter AC.T diff 2TM, electronic analyzer as described in the instruction manual and print outs of haemograms were obtained. These samples were analyzed on the Bio-Rad Variant I HPLC system with the use of the Variant I Beta Thalassaemia short programme Recorder Pack (Bio Rad Laboratiers) as described in the instruction manual. In this system, the samples are mixed on the Variant I sampling station, diluted with the specific hemolyzing wash buffer and injected into an assay-specific analytic cartridge. The separated hemoglobin fractions pass through a flow cell and absorbance is measured at 415 nm while background noise is reduced with the use of a secondary wavelength at 690nm. The raw data are integrated and a chromatogram report is generated.

RESULTS

Table 1: Prevalence of Common Haemoglobino -pathies in different age groups in tribal area

Age (yrs)	Total Number	ß -thalassemia trait (%)	Sickle cell trait (%)
< 17	123847	2053(1.65)	16507(13.32)
17 – 35	43333	849(1.95)	2517(5.8)
> 35	1315	33(2.5)	130(9.89)
Total	168495	2935(1.74)	19154(11.37)

Total numbers of persons screened were 3, 17,539. There were 1, 68,495 persons from tribal areas which include Banaskantha, Baroda, Bharuch, Dahod, Narmada, Panchmahal and Sabarkanha districts. There were 1, 49,044 persons from non-tribal areas which include Ahmedabad, Amreli, Anand, Bhavnagar, Gandhinagar, Jamnagar, Kheda, Mehsana, Kutch, Navasari, Patan, Rajkot, Surat, Porbandar, Surendranagar and Valsad districts. In tribal area, 2935 (1.74%) had ß-thalassemia trait, 19154 (11.37%) had sickle cell trait, 966 (0.57%) had sickle cell disease, 2 (0.0016%) had B- thalassemia major, 1, 30, 595 (77.5 %) were normal and 167 (0.099%) had other haemoglobinopathies like HbD, HbE etc. Table I gives age wise prevalence of sickle cell and ß thalassemia in tribal area. The prevalence of ß-thalassemia trait and sickle cell trait is comparable in females and males. (Table 2)

Table 2: Gender wise prevalence of Common Haemoglobinopathies in tribal area

Gender	Total Number	ß -thalassemia trait (%)	Sickle cell trait (%)
Male	85862	1516 (1.81)	9956 (11.6)
Female	82633	1374 (1.66)	9198 (11.13)
Total	168495	2935 (1.78)	19154 (11.37)

Table 3 gives region wise prevalence of sickle cell and ß thalassemia. The prevalence of ß thalassemia minor is very high in Banaskantha district (4.45%). Banaskantha (5.12 %) and Sabarkantha (3.43%) had low prevalence of sickle cell prevalence.

Table 3: Prevalence of Common Haemoglobino -pathies in tribal districts

Name of District	Number	ß-thalassemia trait (%)	Sickle cell trait (%)
Banaskantha	9489	423(4.45)	486 (5.12)
Baroda	31608	381(1.2)	5018 (15.87)
Bharuch	19373	330(1.7)	2737 (14.12)
Dahod	15975	410(2.57)	2079 (13.1)
Narmada	31966	464(1.45)	5377 (16.82)
Panchmahal	23077	422(1.83)	2189 (9.49)
Sabarkantha	37007	505(1.37)	1268 (3.43)
Total	168495	2935 (1.76)	19154 (11.4)

The total population screened in non-tribal area of Gujarat was 1, 49,044. ß thalassemia minor was seen in 3261 (2.18%). 1604 (1.1%) had sickle cell trait, 23(0.01%) had sickle cell disease, 3(0.002%) were suffering from ß thalassemia major, 236 (0.15%) had other haemoglobinopathies and 144153 (96.7%) were normal. Table 4and 5 mentions age wise and sex

wise distribution of haemoglobinopathies in non-tribal regions respectively.

Table 4: Prevalence of Common Haemoglobino -pathis in different age groups in non-tribal region

Age (yrs)	Total Number	ß -thalassemia trait (%)	Sickle cell trait (%)
< 17	24601	618 (2.51)	242 (0.98)
17 – 35	1,23,924	2633 (2.12)	1353 (1.1)
> 35	519	10 (1.92)	9 (1.74)
Total	1,49,044	3261 (2.18)	1604 (1.1)

Table 5: Gender wise prevalence of Common Haemoglobinopathies in non-tribal area

Gender	Total Number	ß-thalassemia trait (%)	Sickle cell trait (%)
Male	82941	1938 (2.33)	793 (0.95)
Female	66103	1323 (2.00)	811 (1.22)
Total	1,49,044	3261 (2.18)	1604 (1.1)

Table VI gives prevalence of common haemoglobinopathies in non-tribal districts. Kutch had highest prevalence (4.1 %) while Surendranagar had the lowest prevalence (0.77%) of ß thalassemia minor. Surendranagar (3.17 %) had the highest and Kutch (0.08 %) had the lowest prevalence of sickle cell trait.

Table 6: Prevalence of Common Haemoglobinopathies in non-tribal districts

District	Number	ß-Thalassemia trait (%)	Sickle cell trait (%)
Ahmedabad	19825	678 (3.4)	138 (0.7)
Anand	24329	482 (1.98)	273 (1.1)
Gandhinagar	3165	71 (2.24)	30 (0.95)
Bhavnagar	1009	28 (2.77)	10 (0.99)
Amreli	468	14 (2.99)	4 (0.86)
Jamnagar	864	30 (3.47)	5 (0.58)
Kheda	1812	45 (2.48)	7 (0.39)
Kutch	2476	102 (4.1)	2 (0.08)
Navsari	12175	230 (1.89)	328 (2.7)
Mehsana	31161	432 (1.36)	51 (0.16)
Patan	12691	173 (1.36)	20 (0.15)
Rajkot	2284	70 (3.1)	25 (1.1)
Surat	31368	732 (2.33)	596 (1.9)
Porbandar	725	21(2.89)	10 (1.38)
Surendranagar	1042	8 (0.77)	33 (3.17)
Valsad	3625	145 (4)	72 (1.97)
Total	149044	3261 (2.18)	1604(1.1)

Out of total 3, 17,539 persons screened, 6169 had ß thalassemia minor (1.95 %) while 20758 (6.54 %) had sickle cell trait. Sickle cell trait prevalence was high in schedule tribes except in Bamaniyas (6.9 %). Sickle cell was prevalent in all the schedules castes while it was seen in many general castes like barot, Brahmins, patels, parmars, rajputs and rabaris.

DISCUSSION

Hemoglobinopathies are prevalent worldwide, but it is more prevalent in some geographical areas. In India, According to hospital based study, average frequency of sickle cell gene is around 5%. The highest frequency of sickle cell gene in India is reported in Orissa (9%), followed by Assam (8.3%), Madhya Pradesh (7.4%), Uttar Pradesh (7.1%), Tamil Nadu (7.1%) and Gujarat (6.4%).[5,6,7] The prevalence of sickle cell trait (6.54 %, n= 20758) in our study is in concordance with the reported prevalence.

Table 7: Distribution of Sickle cell trait and ß-thalassemia trait in different castes of Gujarat

Caste	Sickle cell trait (%)	ß-thalassemia trait(%)
General Caste		
Barot	1.68	3.5
Brahmin	1.98	3.49
Bhanusali	-	8.1
Bhakta	-	7.93
Bhavsar	-	2.59
Chaudhary	15.63	1.97
Lohana	-	6.5
Patel	3.6	1.43
Parmar	10.4	2.6
Patil	-	4.35
Rajput	7.7	2.9
Rabari	1.03	1.52
SC		
Chamar	10.52	3.49
Chauhan	9.39	-
Harijan	8.1	3.52
Solanki	9.22	2.64
Vankar	8.7	2.94
ST		
Adivasi	15.25	2.42
Bamaniya	6.9	-
Bariya	16.5	1.12
Gamit	17.64	
Koli	16.87	
Nayka	15.38	2.19
Rathva	18.65	
Tadvi	16.95	

In our data, the prevalence of sickle cell trait is 11.36% (3.43%-16.82%) in tribal area, while in non-tribal area; it is 1.1% (0.08%-3.17%). Two tribal districts (sabarkantha: 3.43%, banaskantha: 5.12%) have very low prevalence of sickle cell trait. Rathva (18.65%), Nayka (15.38%), Gamit (17.64%), Tadvi (16.95%), Bariya (16.5%), Adivasi (15.25%) and Chaudhary (15.63%) have higher prevalence of sickle cell trait. The prevalence of sickle cell trait in tribal area is significantly higher in lower age group (13.32 % in < 17 years compared to 5.8 % in 17-35 year age group). This is probably due to survival disadvantage in presence of hemoglobinopathy. Balgir et al also has noted similar trend.[8]

The distribution of beta thalassemia is not uniform in the Indian subcontinent. Though certain communities are identified to have high prevalence, it has been detected in almost every Indian population. The prevalence of beta thalassemia trait varies from 1-17% in different populations of India.[9-14] In our data, the prevalence of thalassemia trait is 1.17% (1.2%-4.45%) in tribal area, while in non-tribal areas it is 2.18% (0.77-4.1%).

Over all prevalence (tribal + non-tribal) of β thalassemia trait is 1.95 % (n=6196). Hypochromic microcytic anemia was observed in 21791(6.86%) cases. Underlying β thalassemia trait could have been missed in this group and investigations for iron deficiency followed by repeat HPLC after iron therapy is needed. Bhanusali (Sindhi) (8.1%) Bhakta (7.93%) and Lohana (6.5%) have high prevalence of β thalassemia trait. Bhakta community resides in Surat and Navsari area and the high prevalence is not previously reported. Higher prevalence of β thalassemia trait is previously reported from Sindhis, Punjabis, Kutchis, Lohanas, Mahars, Neobuddhists, kolis and Agris from Maharasthra, & Gowdas and Lingayats from Karnataka.[5,6] Higher prevalence of β thalassemia trait in tribal districts of banaskantha and Dahod is due to predominance of adivasi and vankar community in these two districts. The prevalence of β thalassemia trait is higher in adivasi (2.42 %) and vankar (2.94 %) community. The higher prevalence of β thalassemia trait in Ahmedabad, kutch and Rajkot is due to predominance of sindhi (Bhanusali) and lohana community in these districts.

It is interesting to note that many non-tribal castes like patel, rabari, brahmins also have presence of sickle genes while many tribal castes like adivasi, nayak and baria have significant presence of β thalassemia genes. Although sickle and β thalassemia are more prevalent in certain castes, they are present in most of the castes screened in our study. As reported previously, prevalence of sickle and β thalassemia trait in general castes and schedule castes is confirmed in our study also .[15] This finding is significant as any screening program to be effective all the castes needs to be screened as sickle cell and thalassemia gene is prevalent in most of the castes and tribal as well as non-tribal regions.

CONCLUSIONS

The prevalence of ß-thalassemia trait and sickle cell trait in Gujarat is 1.95% and 6.54% respectively. In tribal area, 1.74% has ß-thalassemia trait and 11.37% has sickle cell trait while in non-tribal area, ß thalassemia trait is seen in 2.18% and 1.1% has sickle cell trait. Rathva (18.65%), Nayka (15.38%), Gamit (17.64%), Tadvi (16.95%), Bariya (16.5%), Adivasi (15.25%) and Chaudhary (15.63%) communities has higher prevalence of sickle cell trait. ß-thalassemia trait is more prevalent in Sindhi (Bhanusali) (8.1%), Bhakta (7.93%) and Luhana (6.5%) community. Sickle cell trait is very common in schedule castes (8.1-10.52 %) while it is seen in many general castes. ß-thalassemia trait is prevalent in all the castes and communities.

ACKNOWLEDGEMENTS

We thank treasurer, Red Cross; Dr. K.G. Patel and Gujarat government for providing the grant for the screening program, all the staff members of Gujarat Red Cross society and health centers for their active help in carrying out the program and patients who participated in the program.

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