Original Article

Prevalence and hematological profile of β-thalassemia and sickle cell anemia in four communities of Surat city

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BACKGROUND: From the data of transfusion-dependent thalassemia major cases, the 4 communities (Muslim, Dhodia Patel, Kachhiya Patel, and Modh Bania) with high prevalence but not studied methodically were selected.
AIM: The aim of this study is to find prevalence of β-thalassemia and sickle cell anemia in 4 selected communities and also to evaluate hematological profile in them.
MATERIALS AND METHODS: For screening of β-thalassemia trait (BTT) and sickle cell trait (SCT), all samples were tested for red cell indices, solubility, HbA_2 level and doubtful cases confirmed on HPLC.
STATISTICAL ANALYSIS: Mean ± SD, χ^2 and 't' tests were used to evaluate the significance.
RESULTS AND CONCLUSION: Among 4 selected communities, the highest prevalence of BTT was observed in Modh Bania (6.2%) and Kachhiya Patel (6.05%) and that of SCT in Dhodia Patel (14.0%). Significantly higher prevalence of BTT was observed in Memon ($P < 0.0001$) and of SCT in Khalifa 6.6% ($P < 0.0001$) compared to other Muslim sub castes. Anemia was more prevalent in BTT compared to non-BTT and non-SCT subjects. 80% of Dhodia Patel non-BTT and non-SCT subjects showed microcytic red cell morphology. Their Mean ± SD Hb concentration was 12.1 ± 1.73, hence iron deficiency cannot be a sole reason. This community needs α-thalassemia and iron studies.

Key words: β-thalassemia trait, Dhodia Patel, Kachhiya Patel, Modh Bania, Muslim, sickle cell trait, Surat

Introduction

The distribution of β-thalassemia and sickle cell disorders varies geographically and from community to community. Various hemogloginopathies are major public health problem in Gujarat, but the data pertaining to their occurrence and prevalence, especially in Surat, are scarce. According to the census of India Report 2001 (Surat Municipal Corporation), the population of Surat district is 4,995,174; in rural region, it is 1,999,357 and in urban area, it is 2,995,817. Surat has cosmopolitan population, and several communities are living in the district from centuries. Gujarati community dominates and others are Marwaris, Punjabis, Marathi, and Hindi speaking north Indian population. The analysis of transfusion-dependent thalassemia major cases, attending our center, had suggested that there is a high prevalence in Muslims, Patels, Sindhis, Modh Banias, and Mahayavanshi. The sickle cell disease (SCD) was predominantly found in Dhodia Patels, Kolis, and Gamits.

Muslims are divided into 2 major religious, endogamous sects – Shia and Sunni, and several castes like Momins, Memon, Khoja, Bohra, Pathan, Sayied, Shaikh etc. are found in them. As many Muslim sub-castes originate from different geographical locations, this community was selected. This study includes only Sunni Muslims who follow the trend of consanguineous marriages.

Certain sub-castes of Patel[1] and tribal communities are already studied for sickle cell disorders.[2-4] Dhodia Patel, the third largest tribal group in Gujarat, needs β-thalassemia studies as thalassemia major cases are identified in this community. The majority of them are settled in Surat and Valsad districts, but they are also found in Daman and Diu, Dadra and Nagar Haveli, Madhya Pradesh, Maharashtra, Karnataka, Rajasthan states.

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There are no studies on Modh Bania and Kachhia Patel communities of Surat. They originate from town Modhera in Patan district in the northern part of Gujarat. They are predominantly Brahmins or Banias, and those who migrated to Surat are known as Surti Modh Banias or Ghanchi. Majority of Kachhiya Patels are vegetable vendors and cultivators and are basically Kadva Patels. They are inhabitants of Ahmedabad, Surat, Anand, and Khambhat cities in Gujarat.

Materials and Methods

The study was approved by the institutional ethics committee. As a control, 24,917 randomly selected unrelated individuals including all major castes, attending hemoglobinopathies screening camps organized in different locations (school and colleges) in Surat were screened. Total 9,447 samples of selected communities were collected during functions after taking permission from their leaders.

The informed consent in local language was taken, and then, 2 ml blood was collected in EDTA. All the samples were tested for red blood cell indices by an automated cell counter MEK-5216K (NIHON KOHDEN, Japan),[5] solubility test,[6] and cellulose acetate electrophoresis at pH 8.9.[7] The samples showing HbA_2 >3.5% were initially diagnosed as β-thalassemia trait (BTT). The doubtful cases and all traits were confirmed by high performance liquid chromatography (HPLC) on Hemoglobin Variant Testing System, BioRad Laboratories.[8,9] Statistical evaluation of the data was done by mean ± standard deviation (SD), χ^2, and '*t*' tests.

Results

Among 9,447 study participants, 4,870 were Muslims, 2,249 Modh Bania, 1,173 Kachhiya Patel, and 1155 were Dhodia Patel. Sex- and age-wise analysis of the 9,447 subjects showed that 5,385 (57.0%) were male and 4,062 (42.9%) were female having average age 23.0 ± 15.1 years. In control group, 12,970 were male and 11,947 female and had average age of 17.8 ± 2.61 years.

The mean cell volume (MCV) ≤76 fl and mean cell hemoglobin (MCH) ≤26 values suggest BTT.[5] MCV ≤76 fl was observed in 80.0% Dhodia Patel, 33.1% Muslims, 21.8% in Kachhiya Patel, 21.5% Modh Bania, and 16.6% in control population [Table 1]. In Dhodia Patel, incidence of low MCV was significantly higher compared to control population (χ^2 = 72, P < 0.0001). The MCH value of ≤26 value was observed in 80.3% Dhodia Patel, 46.0% in Kachhiya Patel, 39.6% in Muslims, 24.0% in Modh Bania, and 15.5% in Control population. In Dhodia Patel, significantly more subjects were having MCH ≤26 pg compared to control population (χ^2 = 72, P < 0.0001).

The overall prevalence of BTT and SCT in Surat population (control) was 3.2% and 1.38%, respectively [Table 2]. The significantly higher prevalence of BTT was observed in Modh Bania and Kachhia compared to remaining communities and control population of Surat by χ^2-test (P < 0.0001). Dhodia Patels had significantly higher prevalence of SCT (P < 0.0001).

Table 3 shows prevalence of BTT and SCT in different Muslims sub-castes. Significantly higher prevalence of BTT is observed in Memon (χ^2 = 23.8, P < 0.0001) and of SCT in Khalifa (χ^2 = 72, P < 0.0001) compared to other Muslim sub-castes. 4 samples of Muslim community had

Table 1: MCV and MCH values in study population

Parameters	Control		Dhodia Patel		Kachhiya Patel		Modh Bania		Muslims	
	n	%	*n*	%	*n*	%	*n*	%	*n*	%
MCV (fL)										
≤76	4138	16.6	925*	80	256	21.8	485	21.5	1613	33.1
76.1-101	19494	78.2	228	19.7	874	74.5	1722	76.5	3251	66.7
>101	1285	5.1	2	0.2	43	3.6	42	1.8	6	0.12
MCH (pg)										
≤26	3865	15.5	938*	80.3	540	46.0	540	24	1929	39.6
26.1-32	16881	67.7	208	18.0	595	50.7	1460	64.9	2889	59.3
>32	4171	16.7	19	1.6	38	3.2	249	11.0	52	1.06

* Significantly higher incidence of low MCV and MCH compared to other communities and control population of Surat by χ^2-test (P < 0.0001)

Table 2: Prevalence of β-thalassemia trait and sickle cell trait in study population

Community	Total collection	BTT		SCT		SCD	
		n	%	*n*	%	*n*	%
Dhodia Patel	1155	25	2.1	162**	14.0	8	0.69
Kachhia	1173	71*	6.05	2	0.17	0	0
Modh Bania	2249	140*	6.2	5	0.22	0	0
Muslims	4870	139	2.85	59	1.21	2	0.04
Total	9447	375	3.91	228	2.41	10	0.1
Control	24917	809	3.2	343	1.38	11	0.04

* Significantly higher prevalence of BTT in Kachhia and Modh Bania compared to remaining communities and control population of Surat by χ^2-test (P < 0.0001)
** Significantly higher prevalence of SCT in Dhodia Patel compared to remaining communities and control population of Surat by χ^2-test (P < 0.0001)

Table 3: Prevalence of β-thalassemia and sickle cell trait in Muslim sub-castes

Sub Caste	Total collection	BTT *n* (%)	SCT *n*(%)	SCD *n* (%)
Ansari	146	4 (2.7)	0	0
Dawoodi Bohra	704	14 (1.98)	0	0
Fakir	88	5 (5.6)	3 (3.40)	0
Modh Bania	243	9 (3.7)	3 (1.23)	0
Khalifa	121	1 (0.82)	8 (6.6)*	2 (1.65)
Khatki	116	2 (1.72)	3 (2.58)	0
Malek	68	2 (2.94)	0	0
Memon	316	23 (7.2)*	3 (0.94)	0
Momin	95	4 (4.2)	1 (1.05)	0
Pathan	306	8 (2.6)	6 (1.96)	0
Patni	147	4 (2.7)	1 (0.68)	0
Pinjara	130	1 (0.7)	1 (0.76)	0
Sayied	162	1 (0.6)	3 (1.85)	0
Shaikh	1353	35 (2.58)	20 (1.47)	-
Vohra	343	8 (2.33)	4 (1.16)	-
Others	532	18	3	-
Total	4870	139 (2.8)	59 (1.2)	2 (0.04)

Figures in the parenthesis indicate % values* Significantly higher prevalence compared to other Muslim sub-castes by χ^2-test ($P < 0.0001$)

borderline HbA_2 values.

Table 4 shows highly significant ($P < 0.001$) differences in hematological parameters of non-BTT/non-SCT study subjects. In Dhodia Patel, the mean MCV (69.8 ± 8.35) and MCH (23.6 ± 2.03) are significantly lower and RBC (5.24 ± 0.73) values are raised compared to control population. Kachhiya Patels have reduced mean cell hemoglobin concentration (MCHC) (30.82 ± 1.4).

Selected communities and control population showed significantly low values of hemoglobin (Hb) in BTT/SCT subjects compared to non-BTT/non-SCT individuals. As seen in Table 5, anemia is predominant in Kachhiya Patel BTT subjects, and they also have significantly reduced MCH (18.9 ± 2.9), MCHC (29.18 ± 1.61), compared to control population by "*t*" test ($P < 0.001$). Muslim BTT subjects have lowest values of MCV and RDW compared to control population by "*t*" test ($P < 0.001$).

Table 6 shows the significant reduction in MCV, MCH, and Hb S in Dhodia Patel SCT subjects compared to Muslims and control ($P < 0.001$).

Comparison of Hb and HCT in control, BTT, and SCT population shows that anemia is more prevalent in BTT compared to non-BTT/non-SCT subjects.

Discussion

The first step in population screening for thalassemia trait is the accurate complete blood count. As per Dacie and Lewis,[5] MCV ≤76 fL and MCH ≤26 pg indicate possibility of BTT and should be further screened for HbA_2 level. In our study, majority (80%) of Dhodia Patel samples showed reduced MCV and MCH, but cellulose acetate membrane electrophoresis and HPLC confirmed BTT only in 2.1% subjects [Table 2]. Red blood cell indices are also low in α-thalassemia trait and iron deficiency.[10] In India, because of the high prevalence of iron-deficiency anemia, differential diagnosis of BTT is often complicated. Mehta and Pandya[11] have suggested that the BTT individuals have an advantage in maintaining an iron balance. The mean ± SD Hb concentration in non-BTT/non-SCT Dhodia Patel individuals was 12.1 ± 1.73; hence, iron deficiency may not be present in all those having low indices. However, iron deficiency and α-thalassemia studies are essential for this community to explain reasons for low RBC indices.

An increased HbA_2 level (>3.5%) is the hallmark of diagnosis of β-thalassemia carriers. Samples with borderline HbA_2 probably due to silent mutation, co-inheritance of δ and β thalassemia, some mild β-gene mutations, and γδβ-thalassemia should be repeated on HPLC and may be analyzed by molecular methods.[12] In this study, 4 such samples of Muslims community were found, which need to be further confirmed by molecular analysis. Studies in different Indian regions have reported 1% to 17% prevalence of BTT with mean of about 3.3%.[13,14] Comparable prevalence of 3.2% of BTT was found in Surat population selected as control. Madan *et al.*[15] in ICMR multi-center study reported 2.68% BTT prevalence in Mumbai and 5.47% in Delhi. They found 2.7% BTT in Baniya and 2.5% and 1.7% BTT in Sunni Muslims from Mumbai and Delhi, respectively.[15] While in our study, we found 6.2% BTT prevalence in Modh Bania and 2.85% in Muslims. This is the first report on the prevalence of β-thalassemia in Kachhiya Patel community.

Hb AS has been reported as 0% to 31.4% in tribal population of Gujarat.[2] Different non-tribal caste groups are also known to have sickle gene.[16,17] In our study, the prevalence of SCT was 14.0% in Dhodia Patel (tribal community). Earlier studies have reported 13.76% and 17.84% prevalence in this community, in Valsad

Table 4: Hematological data (mean ± S.D.) of non-β-thalassemia trait/non-sickle cell trait selected caste groups and control population

Parameters	Muslims (*n* = 4462)	Dhodia Patel (*n* = 963)	Kachhiya Patel (*n* = 1007)	Modh Bania (*n* = 1292)	Control population (*n* = 23730)
Hb (g/dL)	12.8 ± 3.8*	12.1 ± 1.73*	12.06 ± 1.8*	12.8. ± 1.96*	13.1. ± 1.8
RBC (× 10^6/L)	4.81 ± 0.52*	5.24 ± 0.73*	4.57 ± 0.53	4.59 ± 0.51*	4.63 ± 0.63
HCT (%)	37.9 ± 6.39*	36.4 ± 5.2*	39.8 ± 5.40	37.9 ± 5.23*	39.3 ± 5.5
MCV (fL)	79 ± 7.82*	69.8 ± 8.32*	85.89 ± 9.87	82.7 ± 8.04*	85.5 ± 9.5
MCH (pg)	26.6 ± 3.18*	23.6 ± 3.49*	26.53 ± 3.58*	28.04 ± 3.45*	29.2 ± 3.6
MCHC (g/dL) μ (10^6/μL)	33.56 ± 2.6	33.7 ± 2.89	30.82 ± 1.4*	33.9 ± 3.12	33.6 ± 2.9
RDW (%)	14 ± 2.60*	15.6 ± 2.03*	14.5 ± 1.4*	14.25 ± 1.25*	14.7 ± 1.8
HbA_2 (%)	2.56 ± 0.44	2.67 ± 0.47*	2.5 ± 0.4	2.54 ± 0.43	2.58 ± 0.5

n: Number tested* Statistically significant increase or decrease in mean values by "*t*' test ($P < 0.001$)

Table 5: Hematological data (mean ± S.D.) of β-thalassemia trait subjects

Parameters	Muslims (*n* = 139)	Dhodia Patel (*n* = 25)	Kachhiya Patel (*n* = 71)	Modh Bania (*n* = 140)	Control population (*n* = 809)
Hb (g/dL)	10.9 ± 1.31	11.79 ± 1.67	10.14 ± 1.35*	10.9 ± 1.49	11.6 ± 1.69
RBC (× 10^6/μL)	5.82 ± 0.64	5.42 ± 0.79	5.45 ± 0.74	5.54 ± 0.59	5.46 ± 0.84
HCT (%)	34.71 ± 3.97	35.98 ± 5.54	34.7 ± 4.43	33.9 ± 4.29	35.4 ± 5.3
MCV (fL)	59.77 ± 5.38*	64.3 ± 6.6	64.19 ± 7.22	61.2 ± 4.7*	65.1 ± 6.6
MCH (pg)	18.95 ± 2.36	21.56 ± 3.01	18.9 ± 2.9*	19.7 ± 2.11	21.6 ± 3.07
MCHC (g/dL)	31.48 ± 2.52*	33.45 ± 2.58	29.18 ± 1.61*	32.2 ± 2.79	33.1 ± 3.3
RDW (%)	15.31 ± 1.35*	16.54 ± 1.52	16.2 ± 1.7	15.8 ± 1.4	15.8 ± 1.7
HbA_2 (%)	5.32 ± 0.82	5.8 ± 0.5	4.96 ± 0.6	5.17 ± 0.75	5.1 ± 1.0

n: Number tested* Statistically significant increase or decrease in mean values by "*t*' test ($P < 0.001$)

Table 6: Hematological data (mean ± SD) of sickle cell trait subjects

Parameters	Muslims (*n* = 59)	Dhodia Patel (*n* = 162)	Control population (*n* = 343)
Hemoglobin (g/dL)	13.09 ± 1.9	12.2 ± 1.5	12.4 ± 1.9
RBC count (× 10^6/L)	5.02 ± 0.6	5.34 ± 0.7	5.00 ± 0.8
HCT (%)	37.5 ± 4.3	36.4 ± 4.5	37.6 ± 5.9
MCV (fL) μ (10^6/μL)	75.3 ± 7.8	68.6 ± 7.8*	75.1 ± 11.4
MCH (pg)	25.8 ± 2.4	23 .0 ± 2.77*	25.1 ± 4.6
MCHC (g/dL)	34.9 ± 2.7	33.5 ± 2.8	33.0 ± 3.07
RDW (%)	13.9 ± 1.0*	15.5 ± 2.2	15.8 ± 2.3
HbA_2 (%)	3.1 ± 0.5	3.4 ± 0.6*	2.9 ± 0.8
Hb S (%)	34.02 ± 4.2	27.9 ± 3.4*	31.2 ± 5.8

*Statistically significant increase or decrease in mean values by "*t*' test ($P < 0.001$)

and Surat respectively.[2] Mukherjee *et al.*[3] reported the lower Hb S level (27.9%) in tribal compared to the non –tribal group (35.5%). They also reported very high frequency of α-thalassemia gene in tribal (0.97) compared to the non-tribal (0.24). We found similar results in Dhodia Patel with reduced Hb S level (27.9 ± 3.4) compared to Muslims (34.02 ± 4.2) and control (31.2 ± 5.8). Very high incidence of reduced RBC indices in Dhodia Patel indicates the possibility of α-thalassemia gene in this tribe.

Compared to normal (negative for BTT or SCT) subjects, Hb and hematocrit values were significantly lower in BTT subjects. The ICMR study[15] has also detected more anemic children in BTT series compared to children without any hemoglobinopathy.

Conclusion

BTT prevalence is higher in Modh Bania and Kachhiya Patel, and higher SCT prevalence is observed in Dhodia Patel, Memon, and Khalifa. As majority of Dhodia Patels show microcytic red cell morphology, this community also needs α-thalassemia and iron study.

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