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Sickle cell anaemia & glucose-6-phosphate dehydrogenase (G6PD) deficiency in Lahaul & Spiti, a tribal district of Himachal Pradesh

Inherited haemoglobin disorders (thalassaemia and sickle cell disease) primarily affect the red blood cells and are more commonly seen in certain geographical areas and ethnic groups[1,2]. When untreated, these disorders cause severe morbidity and mortality in addition to the extreme socio-economic burden on the family, society and health services. With the advances in medical sciences and the development of health resources, it is now possible to provide necessary care, prevention and control of these disorders. The government of India has recently initiated a nationwide programme for providing adequate care, prevention and management of various genetic disorders[3]. Following the Government of India directive, several States initiated community-based screening programmes[4].

The screening was carried out for sickle cell anaemia and glucose-6-phosphate dehydrogenase (G6PD) deficiencies in Lahaul and Spiti by ICMR-National Institute of Research in Tribal Health Field Station, Keylong on the request of the Himachal Pradesh State government. Lahaul and Spiti is the tribal-dominated district of Himachal Pradesh lying on the Indo-Tibetan border. Lahaul and Spiti are the two far-flung Himalayan valleys of Himachal Pradesh covering an area of 13,835 km^2 with snowfields, ice caves, glaciers and snow-clad mountains and valleys. Lahaul valley is situated towards the West with Keylong as its headquarters whereas; Spiti valley lies in the middle of Ladakh and Tibet and on the Eastern side of the district with Kaza as its headquarters. It is a mountainous district of Himachal Pradesh having altitude varying from 3000 to 4500 m. Community-based screening is a challenging task under given the adverse climatic conditions in these locations.

A prospective community-based cross-sectional study was done wherein a total of 2809 individuals of all age groups and both genders were randomly selected from 65 (49 from Lahaul and 16 from Spiti) villages from Tinnan, Pattan, Gahar, Todd valleys in Lahaul area and Spiti valley from May 2016 to November 2018. Sample size was calculated assuming 10 per cent prevalence, and 95 per cent level of confidence, 2.5 per cent absolute error and 32,000 total population, a minimum sample required for a cross-sectional study was 544. Since, a random sample was not possible in such areas, where a population of 32,000 is spread over 13,833 km^2 with population density of about 2 persons per sq μm, a design effect of two was assumed and 10 per cent refusal rate, so, 1197≈1200 samples were required. To have a comparative study, the equal number of males and females were sampled (1200 each; total sample=2400). Complete demographic details of all the individuals were recorded. Written informed consent was taken from all the individuals/parents of participants. Two millilitre of intravenous blood was drawn in an EDTA vial under aseptic conditions from all the individuals at the field site. The samples were stored at 4°C and transported to the laboratory. To screen for sickle cell anaemia, solubility test[5] was performed. Confirmation of genotype and for beta-thalassaemia and other haemoglobin variants, cellulose acetate electrophoresis and CE-HPLC (High Performance Liquid Chromatography) was performed. G6PD deficiency was checked for using the standard dichlorophenol indophenol decolourization method[6]. Data were analyzed with the continuous variables presented as median (25^{th}-75^{th} centile), and the discrete variables presented as numbers (percentages).

Out of the 2809 individuals tested, 1367 (48.7%) were male and 1442 (51.3%) were female (Table). The median age was 37 yr (17-52 yr). Table depicts the community, age and valley-wise breakup of the participants. Bodh (78.7%) and Swangla (11.7%) are two tribal groups mainly inhabiting the district. The

Table. Age and gender wise distribution of studied cohort in Lahaul and Spiti

Age group	Spiti*		Lahaul#						Total
	Bodh		Bodh		Swangla		Others		
	Male	Female	Male	Female	Male	Female	Male	Female	
0-10	35	17	23	24	3	5	9	8	124
11-20	111	70	72	90	87	106	57	71	664
21-30	81	45	82	90	16	17	49	18	398
31-40	59	65	116	128	19	15	24	12	438
41-50	44	49	130	169	12	15	10	3	432
51-60	45	26	120	192	5	12	6	0	406
61-70	19	8	72	113	3	4	2	0	221
>70	7	4	42	63	7	3	0	0	126
Total@	401 (a1)	284 (b1)	657 (a2)	869 (b2)	152 (a3)	177 (b3)	157 (a4)	112 (b4)	2809
	685 (T1)		1526 (T2)		329		269		2809
Community-wise	Bodh (T1+T2)				Swangla		Others		
Total	2211 (78.7%)				329 (11.7%)		269 (9.57%)		

@Male: a1+a2+a3+a4=1367 (48.7%); Female: b1+b2+b3+b4=1442 (51.3%); *Majority of the population is of Bodh category in Spiti area while swangla and other non-tribal communities were almost negligible, hence not screened; #The population of Lahaul valley comprises Bodh category in majority followed by swangla and other non-tribal communities including migrant workers, employees, *etc.*

Swangla tribes are a small group of Hindus and their main occupations are agriculture and animal husbandry. They generally speak the Tinani language and are mainly present in the Pattan valley while Bodh tribal community is predominant in Tinnan, Gahar and Todd sub-valleys. Bodhs are also known as 'Bhotia' and their preferred religion is Buddhism. The remaining 9.57 per cent of individuals belonged to other categories with a majority of these being migrant workers from other districts. None of the 2809 individuals tested, showed the presence of HbS or beta-thalassaemia trait. Similarly, no G6PD deficiency was detected. Efforts are, ongoing to screen for these haemoglobinopathies in lower altitudes of the State.

Sickle cell disease is common among tribes of India and its prevalence ranges from one to 40 per cent[2]. The disease is generally milder among Indian patients with fewer episodes of painful crises, infections, acute chest syndrome and need for hospitalization. The severity of the disease increases with exposure to extreme climate conditions leading to high morbidity and mortality[7]. Even sickle cell trait which is considered a Benin condition can develop splenic syndrome at high altitudes which requires medical management[8]. Earlier, O'Brien *et al*[9] had reported infarction of spleen in sickle heterozygote above 2130 altitude. The inhabitants of Lahaul and Spiti valleys are exposed to extreme cold in the winter months and low oxygen pressure due to high altitude.

In the present study, no sickle haemoglobin-positive individual was detected among Bodhs or Swangla tribes of Lahaul and Spiti valley. Bhasin[10] in 2006 reviewed the frequency of abnormal haemoglobins and G6PD deficiency in many tribes and linguistic groups of India and reported the zero per cent frequency of sickle cell gene in the Bhotia (Tibetan group) group. This is similar to our study as Bodh tribes are considered as a sub-tribe of the Bhotia tribe.

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References

1. Mukherjee MB, Colah RB, Martin S, Ghosh K. Glucose-6-phosphate dehydrogenase (G6PD) deficiency among tribal populations of India – Country scenario. *Indian J Med Res* 2015; *141* : 516-20.
2. Colah RB, Mukherjee MB, Martin S, Ghosh K. Sickle cell disease in tribal populations in India. *Indian J Med Res* 2015; *141* : 509-15.
3. National Health Mission. Ministry of Health and Family Welfare. *Government of India. Guidelines on prevention and control of hemoglobinpathies in India*. Available from: *http://www.nhm.gov.in/images/pdf/in-focus/NHM_Guidelines_on_Hemoglobinopathies_in_India.pdf*, accessed on March 5, 2019.
4. Ministry of Tribal Affairs, Government of India. *Swasthya Portal*. Available from: *https://tribal.nic.in/sickle-cell-disease-piramal-swasthya.aspx*, accessed on March 9, 2019.
5. Chanarin I. Laboratory Haematology: An account of laboratory techniques.1st ed. London: Churchill Livingstone; 1989.
6. Bernstein RE. A rapid screening dye test for the detection of glucose-6-phosphate dehydrogenase deficiency in red cells. *Nature* 1962; *194* : 192-3.
7. Tewari S, Brousse V, Piel FB, Menzel S, Rees DC. Environmental determinants of severity in sickle cell disease. *Haematologica* 2015; *100* : 1108-16.
8. Fernando C, Mendis S, Upasena AP, Costa YJ, Williams HS, Moratuwagama D. Splenic syndrome in a young man at high altitude with undetected sickle cell trait. *J Patient Exp* 2018; *5* : 153-5.
9. O'Brien RT, Pearson HA, Godley JA, Spencer RP. Splenic infarct and sickle-(cell) trait. *N Engl J Med* 1972; *287* : 720.
10. Bhasin MK. Genetics of castes and tribes of India: Glucose-6-phosphate dehydrogenase deficiency and abnormal haemoglobins (HbS and HbE). *Int J Hum Genet* 2006; *6* : 49-72.