Spectrum of Hemoglobinopathies in Eastern Uttar Pradesh

Sir,

The rising prevalence of hemoglobinopathies worldwide[1] is of special significance in developing countries like India due to an increased burden on health care delivery system[2]. A number of studies from different parts of the country provide the profile and ethnic distribution of various hemoglobinopathies[3, 4] but there is limited data from Eastern Uttar Pradesh. The present study was undertaken to study the spectrum, ethnic and geographic distribution of hemoglobinopathies in this region.

The study was carried out over a period of 4 years in the Thalassemia Unit of a tertiary care referral hospital catering to a large population of Eastern Uttar Pradesh and adjoining districts of Bihar. Therefore, the data collected is representative of this region. Background data of each index case like age, sex, caste, place of origin and hematological investigations including red cell indices, examination of peripheral smear and hemoglobin electrophoresis were recorded in all cases. For ethnic distribution, patients were divided into 4 categories according to their castes, prevalent in the region. Category 1: higher castes (Brahmin, Kshatriya, Bhoomihar, Kayastha, Vaishya), Category 2: Backward and Scheduled castes (Yadav, Kurmi, Kharwar, Teli, Patel, Kumhar *etc*.), Category 3: Muslims and Category 4: Punjabi, Khatri and Sindhi community. The last category was kept separate because these communities are not natives of this region.

A total of 94 cases in the age group of 3 mo to 12 yr were diagnosed as thalassemia and related hemoglobinopathies. The male to female ratio was 3.5:1. Distribution of hemoglobinopathies is shown in table 1. The commonest hemoglobinopathy was homozygous β-thalassemia (78.7%) followed by E-beta- thalassemia (11.7%), sickle cell-β-thalassemia (3.3%), homozygous sickle cell disease (3.2%), beta- thalassemia trait (2.1%) and hemoglobin H disease (1.1%). 67 patients(71.3%) were transfusion dependent thalassemia major, majority of whom were homozygous beta- thalassemia. 22 patients (23.4%) were categorized as thalassemia intermedia which included beta-thalassemia (45.5%), E-beta thalassemia (36.4%) and sickle- beta thalassemia (13.6%). 52.1% patients belonged to backward castes and together with higher castes contributed to 86.1% of patients. 8.5% cases were seen in Muslim patients whereas Punjabi and Sindhi communities contributed only 5.3% of the patients.

Approximately half of the patients were natives of Varanasi and neighbouring districts. One fourth of the patients were from adjoining districts of Bihar and 12.8% were from Mirzapur, Sonebhadra and adjacent districts of Madhya Pradesh and Chhattisgarh which has a sizeable tribal population.

TABLE 1. Spectrum of Hemoglobinopathies and Their Clinical Phenotypes

Clinical Phenotype	Total		Thalassemia Major		Thalassemia Intermedia		Thalassemia Minor	
Hemoglobinopathies	N	%	N	%	N	%	N	%
β/β thalassemia	74	78.7	64	95.5	10	45.5	-	-
HbE/β thalassemia	11	11.7	3	4.5	8	36.4	-	-
HbS/β thalassemia	3	3.2	-	-	3	13.6	-	-
HbS/S disease	3	3.2	-	-	-	-	-	-
HbBeta/A thalassemia	2	2.1	-	-	-	-	2	2.1
HbH disease	1	1.1	-	-	1	1.1	-	-

To conclude, beta- thalassemia was the commonest hemoglobinopathy. Majority of the patients were natives of this region. The disease was not limited to specific community and was present in all the castes of this region.

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