HPLC analysis of Sickle cell disorders and role of fetal hemoglobin on various manifestations of the disease

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Objectives

Recognizing the prevalence of sickle cell anemia in Yavatmal district, the study was carried out to find out distribution of the disorder in Yavatmal and to study the extent of various manifestations in homozygous sickle cell cases & correlate them with fetal hemoglobin levels using HPLC.

Methods

The patients attending OPD and IPD were screened for sickling test, hemoglobin electrophoresis and high performance liquid chromatography(HPLC). So 67 subjects were selected. All the subjects were categorized into sickle cell trait(AS), sickle cell disease(SS) and heterozygous sickle with â-thalassemia. Detailed history of subjects was taken as per the prestructured proforma.

Results

out of 12 different communities studied, highest incidence of disease was found in Mahar community(43.3%). The commonest clinical manifestations in AS subjects were joint pain & abdominal pain whereas in SS subjects, it was joint pain, fever, jaundice and chest pain. Major reasons of hospitalization in SS patients were vaso-occlusive crisis (26.9%), infections (28.4%) and blood transfusion (26.9%). Splenomegaly was found in 4 subjects(SS) of age 20-30 years. Mean HbF% in AS subjects was 1.4±1.36 and in SS it was 20.1±5.78. Mean HbS% in sickle cell trait was 34.6±5.0. Coexistent alpha thalassemia was suspected in 51.72% of AS subjects.

Conclusion

Higher HbF% was found to have protective effect on incidence of convulsions and blood transfusion. However, higher HbF% was not found to ameliorate the incidence of following manifestations like jaundice, pain crisis, headache, infections. So we conclude that fetal hemoglobin plays a variable role in various manifestations of homozygous sickle cell disease. Also suggest that various modulators at genetic level which modify HbF expression in sickle cell patients of Saudi-Indian haplotype (mainly found in India) should be studied.