Brief Reports

Hemoglobin H Disease–A Report of Five Cases

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Alpha thalassemias, the inherited disorders of hemoglobin synthesis with impaired production of alpha chains, are uncommon in our country(1) and very few cases of hemoglobin H (Hb H) disease have been reported in Indian literature(1-5). We report here the clinical and hematologic features of five cases of Hb H disease.

Case Reports

The clinical and hematological parameters of the five cases are tabulated in *Table I*. In all cases peripheral smear examination pointed to a hemolytic anemia. Reticulocyte preparation with 2% Brilliant Cresyl Blue showed dot like Hb H inclusions. The hemolysates were prepared carefully(6) and electrophoresis at pH 8.6 showed a fast moving Hb H band.

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Discussion

Thalassemia syndromes are the commonest hereditary hemolytic anemias. Normal individuals have the genotype. $\alpha\alpha/\alpha\alpha$. One inherits alpha thalassemia due to impaired production of alpha chains. Hb H disease occurs when this production is greatly reduced. This can be due to deletion genotypes, where there is a deletion in both genotypes (--/-α) or to non-deletion genotypes where at least one of the haplotypes does not have a deletion defect (--/$\alpha\alpha$T)(7). Alpha thalassemia is particularly common in South East Asia and the Mediterranean(8) and relatively uncommon in our country(9). In a study of 850 patients by Dash and Das only two cases of Hb H disease were diagnosed(1).

Hb H disease is a chronic hemolytic disorder with considerable variation in severity(4). It has been seen in a wide variety of age groups in India(1,5). Four of our cases were children. All our cases had hemoglobin less than 8 g/dl which is consistent with most reports(1-5). Our first case presented with a hemolytic crisis. Intake of sulfonamide and other drugs have been known to aggravate hemolysis in Hb H disease(2,4). Peripheral blood film examination in all cases revealed moderate anisopoikilocytosis with microcytosis, hypochromia and presence of target cells. A careful evaluation of the supravital dye stained smears reveals dot like Hb H inclusions which need to be differentiated from fine reticulum of reticulocytes. While pre-

TABLE I–*Clinical and Hematologic Parameters in Five Cases of Hb H Disease*

Parameter Age/Sex	Case 1 2/M	Case 2 4/M	Case 3 23/F	Case 4 11/F	Case 5 9/F
Presenting complaints	Unconscious following anti-malarial therapy	Sibling of case 1 asymptomatic	Generalized ill health 10 yrs. recurrent jaundice in the past	h/o fever 15 days past h/o. icterus +	Sibling of case 4 h/o generalized ill health & pallor
Pallor	+ +	+	+ +	+ +	+
Icterus	+	-	-	-	-
Hepatomegaly (cm)	4	-	-	2	2
Splenomegaly (cm)	6	2	2	2	2
Hb (g/dl)	4.2	7.7	6.5	4.8	6.4
PCV (%)	15	23	20	17	20
Reticulocyte (%)	6	4.5	6	5	3
HbH inclusion	+	+	+	+	+
RBC morphology	Abnormal*	Abnormal*	Abnormal*	Abnormal*	Abnormal*
HbF (%)	1.5	2.3	1.5	1.0	0.8
Bone marrow	Not done	Not done	Erythroid hyperplasia iron stores–Gd. II	Not done	Not done

* Moderate anisopoikilocytosis
moderate hypochromia
microcytosis +
tear drop cell +, target cell +
occasional nucleated RBC.

paring the hemolysate for starch agarose electrophoresis at pH 8.6, care has to be taken that the lysates are not shaken too violently with organic solvents as this would precipitate out the Hb H(6,10) and no Hb H band would be seen on electrophoresis, and the diagnosis of Hb H disease may be missed as had happened in case 3 on an earlier occasion. These two simple tests can clinch the diagnosis.

However, further studies like globin chain analysis(5) and antenatal diagnosis(11) and the awareness about the prevalence of this hemolytic disorder would help in diagnosing more cases of alpha thalassemia and in genetic counselling. A thrust in this direction by various scientific and voluntary agencies would go a long way in bringing down the birth rate of α and β thalassemics.

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Screening for Dental Diseases

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Dental diseases including dental caries and periodontal diseases are very common in children. The incidence of these conditions, however, vary from country to country and even in different parts of the same country. The present study was done to assess the oral health status of school children of Rohtak, Haryana.

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