Clinical and pathological status of haemoglobinopathies among pregnant women in southern Orissa

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Sixty-two pregnant women were categorized into four age groups and investigated to know the prevalence of haemoglobinopathies among them in and around Berhampur using sickling test, naked eye single tube red cell osmotic fragility test (NESTROFT), and haemoglobin electrophoresis. Out of the 62 pregnant women four cases of sickle cell trait and one of β-thalassaemia trait was found. There was no significant difference recorded in the blood cell indices between normal and sickle cell trait in pregnant women. Sickle cell haemoglobinopathy is prevalent among the general, scheduled caste followed by other backward class groups of southern Orissa and less HbS% was observed among the pregnant women which indicate the probable interaction of sickle hemoglobin with α-thalassemia.

Keywords: Electrophoresis haemoglobinopathies, sickle cell, NESTROFT

Anemia in pregnancy is emerging as one of the most important causes of maternal morbidity and mortality in many developing countries. More than 50% of all the pregnant women are reported to be anemic in India. Iron (15-80%) and Folate (2-50%) deficiencies are the common causes of anemia in pregnant women1. In India, hereditary hemolytic disorders such as hemoglobinopathies and glucose 6 phosphate dehydrogenase deficiency are additional factors that lead to anemia during pregnancy. Studies on hemoglobinopathies have shown the prevalence of sickle cell gene spread in India2; in the neighboring state of West Bengal, the prevalence rate was found to be 20.47% with age group between 0-9 years3. Hemoglobinopathies and thalassaemia are autosomal recessive inherited disorders primarily involve globin moiety of the hemoglobin molecule and are characterized by the production of structurally defective hemoglobin molecule such as Hbs S, C, D, E, etc. The thalassaemias are characterized by reduced rate of production of normal Hb due to absence or decrease in the synthesis of one or more types of polypeptide chains (α & β). Underproduction of hemoglobin and imbalanced globin chain synthesis leads to a shortened red cell survival rate.

Sixty-two pregnant women, who attended the OPD of M K C G Medical College and Hospital, Berhampur, were investigated during various months of gestation. Measurement of blood cell indices, mean cell volume, mean cell hemoglobin, mean cell hemoglobin concentration, hematocrit, red cell width was done with MS4 electronic cell counter. The diagnosis of abnormal Hb was established by sickling test performed with freshly prepared 2% sodium meta bisulphite and keeping the wet smear sealed for about 3-4 h followed by examination. All the blood samples were subjected to naked eye single tube red cell osmotic fragility test (NESTROFT) for screening of β-thalassaemia. Haemoglobin electrophoresis was done on cellulose acetate paper to find out the type of haemoglobin (SS, AS or AA) with TBE buffer at alkaline pH of about 8.6 (Fig. 1). Fetal haemoglobin was estimated by alkali denaturation method3.

All the 62 pregnant women who formed the study group were between 15-35 years of age. There were 5 patients with hemoglobinopathy such as 4 sickle cell trait (Hb-AS) and 1 β-thalassaemia trait. There was no significant difference observed among the pregnant women in routine hematological indices (Table 1). The HbA2 level was found 4.5% in β-thalassaemia trait subjects in comparison to medical reference value of 1.0-3.5% in normal subjects (Table 2). The study shows the prevalence of sickle cell disorder in scheduled caste (SC) and other backward caste (OBC) groups of southern Orissa (Table 3).

The present study shows the prevalence of about 8% hemoglobinopathies among pregnant women sampled from OPD of M K C G Medical College and Hospital, Berhampur. Out of the total 8%, 6.5% had sickle cell disorder and 1.5% β-thalassaemia. Whereas studies on pregnant women showed 6.7% sickle cell and 1.5% β-thalassaemia out of their total 8.5%4.

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Previous studies reported that for sickle cell trait with double α-gene deletion had lower HbS% i.e less than 28%\(^5\). The present study shows the percentage of sickle Hb is lower i.e. less than 30% for sickle cell trait, which indicates the possible interaction of sickle Hb gene with α-thalassaemia. Due to association of α-thalassaemia there was no α globin chain to combine with sickle β so the HbS% was found to be less than the normal. Majority of the normal pregnant women were under mild grade of anemia. Out of 4 sickle cell trait subjects, 2 were severe grade and 2 were normal grade. It shows that during pregnancy anemic condition is natural either due to any inherited genetic disorder or due to nutritional deficiency. On the basis of the findings of the study it is very important to take an early step towards medication and awareness because the southern Orissa social system becomes more vulnerable to these genetic disorders as the congenial marriages are very common in the society.

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**References**