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Review Article

THALASSEMIA: A REVIEW

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ABSTRACT:-

The thalassemia is a group of inherited hematologic disorders caused by defects in the synthesis of one or more of the hemoglobin chains. Alpha thalassemia is caused by reduced or absent synthesis of alpha globin chains, and beta thalassemia is caused by reduced or absent synthesis of beta globin chains. Imbalances of globin chains cause hemolysis and impair erythropoiesis. Silent carriers of alpha thalassemia and persons with alpha or beta thalassemia trait are asymptomatic and require no treatment. Alpha thalassemia intermedia, or hemoglobin H disease, causes hemolytic anemia. Alpha thalassemia major with hemoglobin Bart's usually results in fatal hydropsfetalis. Beta thalassemia major causes hemolytic anemia, poor growth, and skeletal abnormalities during infancy. Affected children will require regular lifelong blood transfusions. Beta thalassemia intermedia is less severe than beta thalassemia major and may require episodic blood transfusions. β-Thalassemia and sickle cell disease both display a great deal of phenotypic heterogeneity, despite being generally thought of as simple Mendelian diseases. The reasons for this are not well understood, although the level of fetal hemoglobin (HbF) is one well characterized ameliorating factor in both of these conditions. The globin gene disorders including the thalassemias are among the most common human genetic diseases with more than 300,000 severely affected individuals born throughout the world every year. Because of the easy accessibility of purified, highly specialized, mature erythroid cells from peripheral blood, the hemoglobinopathies were among the first tractable human molecular diseases.

Key words:- *Alpha thalassemia, Beta thalassemia, Hemoglobin*