Introduction

Thalassemia syndromes are the most commonly genetically determined disorders of hemoglobin (Hb) biosynthesis.4.83 percent of the world's population carries globin variants, including 1.67 percent of the population who are heterozygous for α-thalassemia and β-thalassemia. In addition, 1.92 percent carry sickle hemoglobin, 0.95 percent carry hemoglobin E, and 0.29 percent carry hemoglobin C. Thus, the worldwide birth rate of people who are homozygous or compound heterozygous for symptomatic globin disorders, including α-thalassemia and β-thalassemia, is no less than 2.4 per 1000 births, of which 1.96 have sickle cell disease and 0.44 have thalassemias ***(Angastiniotis et al ., 1998).***

Beta thalassemia major was first described by Cooley in 1925 in Greece. ***(Lukens JN et al., 1999)***

Thalassemia syndromes are a heterogeneous group of inherited anemias characterized by defects in the synthesis of one or more of globin chain subunits of hemoglobin (Hb) tetramer .The clinical syndromes associated with thalassemias arise from the combined consequences of Hb production and unbalanced accumulation of globin subunits. The former causes hypochromia and microcytosis.The later leads to ineffective erythropoiesis ***(Forget et al., 2000).***

New developments in the epidemiology, treatment and prognosis of thalassemia have dramatically altered the approach to the care of affected patients, and those developments are likely to have an even greater impact in the next few years.Improved survival of patients with beta thalassemia has given new importance to adult complications such as endocrinopathies, hepatitis and cardiac complications which have a major impact on the quality of life. ***(Cohen AR et al., 2004).***

Conclusion

β-thalassemia is a significant public health problem in Egypt where over 1000 of the annual 1.5 million newborns are expected to be affected with this disorder ***(Rady et al., 1997).***

The removal of the spleen stabilizes the hemoglobin concentration at higher levels and reduces transfusion needs. This may contribute to the prevention of additional cardiac deterioration by limiting myocardial hypoxia and iron overload.

Regular transfusion and iron chelation therapy have increased the duration and quality of life in patients with beta thalassemia major. Heart failure and hepatic insufficiency are the most common causes of death in these patients, but with modern treatment and long survival, endocrine dysfunction assumes greater importance ***(Beris, 1995).***

But splenectomy increases the risk of development of PHT. These results may be useful for the design of therapeutic interventions , since early application of intensive therapy in TM have been shown to prevent the complications related to chronic anemia.