

INTRODUCTION

The thalassemias are heterogenous group of genetic disorders of hemoglobin synthesis. All of which result from reduced rate of production of one or more of globin chains of hemoglobin. They are divided into α - β thalassemia according to defect in globin chain synthesis (*Aessopos et al., 2001*).

Primary management of patients with thalassemia major can be considered under 3 major heading, transfusion therapy, iron chelation therapy and splenectomy (*Thuret, 2001*).

Regular blood transfusion eliminates the complications of anemia (*Weatherall and Clegg, 1995*). However, transfusion results in a "second disease" while treating the first, that of unavoidable accumulation of tissues iron that, without treatment, is fatal in the second decade of life (*Cohen, 1987*).

Thalassemia has many complication as affected infants fail to thrive. Some children show retarded growth early in life, although in general, slowing of the growth is more marked as puberty approaches (*de Sanctis, 2002*). Primary amenorrhea in females, delayed puberty in males caused by chronic anaemia and endocrine disturbance (*Lanzkowsky, 2000*).

Desferrioxamine is an iron chelating agent, has been the only chelating agent in widespread use. This drug dramatically alters the prognosis of this previously fetal disease (*Olivirie et al., 1997*).