

INTRODUCTION AND HISTORICAL BACKGROUND

Thalassemia is a form of hereditary hemolytic anemia characterized by decreased or absent synthesis of one of the globulin subunits of the hemoglobin molecules [Weatherall and Clegg, 1981].

It was first described by Dr. Thomas B. Cooley and Colleagues in Detroit [Cooley and Lee, 1925]. The term "thalassemia", derived from the Greek "θαλασσα" (the sea), was first coined by Whipple and Bradford in 1936 to indicate the association of the disease with the Mediterranean area. It was only since 1940 that the true genetic character of this disorder was fully appreciated by a number of workers in Europe and the United states independently; these included Silvestroni and Bianco, Wintrobe and coworkers, Damashek, Strauss and coworkers, Valentine and Neel, Gatto, and Smith [Weatherall and Clegg, 1981].

The treatment goals for patients with thalassemia major have undergone a radical change during the last years. The combination of iron chelation treatment with the maintenance of hemoglobin levels in a higher range has been shown not only to improve the quality of life but also to prolong survival [Matthews, 1988].