Thesis Title	A Study on the Thiamin Status in β-Thalassemia/Hb E Disease
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ABSTRACT

The β-thalassemia/Hb E is a hereditary hemolytic anemia characterized as the compound heterozygote between β-thalassemic and Hb E. The clinical picture of the patients include defective physical development, mongoloid or thalassemic facies, anemia hepatosplenomegaly and heart failure. Heart disease is a problem in thalassemia and the patients are reported to die from cardiac dysfunction. Abnormal cardiac function, especially congestive heart failure may occur in subjects with thiamin deficiency.

In this study we evaluated the thiamin status in normal subjects and patients with β-thalassemia/Hb E disease. The mean red cell thiamin levels were 8.33 $\pm$ 4.51, 7.40 $\pm$ 3.50, 7.16 $\pm$ 5.09 ug/10^{12} RBCS in normal subjects (n=10), nonsplenectomized β-thalassemia/Hb E (n=13) and splenectomized β-thalassemia/Hb E (n=11),

respectively. The mean red cell thiamin in thalassemic patients was not significantly different from the normal subjects ($p > .05$). The transketolase activity in normal subjects (n=32), nonsplenectomized β-thalassemia/Hb E (n=36) and splenectomized β-thalassemia/Hb E (n=32) were 72.04 ± 14.21,102.11 ± 18.44 and 129.00 ± 18.44 IU, respectively.The mean transketolase activity of these patients was significantly higher than those of the normal subjects ($p<0.05$), and the splenectomized β-thalassemia/Hb E had significantly higher transketolase activity than that of the nonsplenectomized β-thalassemia/Hb E. However, the range of TPP effect in the patients with and without splenectomy are in the range of normal subjects [normal subjects (n=32) 13.14 ± 3.30 %, nonsplenectomized β-thalassemia/Hb E (n=36) = 6.80 ± 5.09 % and splenectomized β-thalassemia/Hb E (n=32) = 4.56 ± 2.63 %). These findings suggested that there was no evidence of thiamin deficiency in patients with thalassemia in the present study.