The Frequency of Adrenal Insufficiency in Adolescents and Young Adults with Thalassemia Major versus Thalassemia Intermedia in Iran.

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Abstract

BACKGROUND:
Endocrine dysfunction is not uncommon complication in patients with transfusion-dependent thalassemia and is thought to occur as a consequence of excessive iron overload. The primary objective of this study is to determine the frequency of adrenal insufficiency in patients with thalassemia major and thalassemia intermediate.

METHODS:
This cross-sectional study was done at the Shiraz University of Medical Sciences, Shiraz, Southern Iran, in 2013. One hundred and ninety patients were divided into two groups; thalassemia major(TM) and thalassemia intermediate (TI) groups. We measured 8 AM serum cortisol, ACTH and ferritin concentrations in all patients.

RESULTS:
The mean age of the TM and TI group were 22.5±5.7 and 23.8±6 years, respectively. 90 patients (47.4%) were splenectomized, 34 (36.2%) with TM and 56 (58.2%) with TI (p:<0.001). The median and interquartile range of serum ferritin levels were 2184±3700 ng/ml and 437±443ng/ml in TM and TI respectively (p< 0.001). Three patients with TM (1.6%) had low basal cortisol and ACTH levels. However, their cortisol response to standard dose ACTH was normal in all patients with TM and TI.

CONCLUSIONS:
Low basal concentrations of cortisol and ACTH occurred in 1.6% of our adolescents young adult patients with TM suggesting a central defect in cortisol secretion at the basal state. However, cortisol response to standard - dose ACTH was normal in all patients with TM and TI.