

SUMMARY

Introduction and aim: The most important reason of morbidity and mortality in patient with thalassemia major (TM) and thalassemia intermedia (TI) is secondary iron storage. While the main source of the iron storage in the body is the blood transfusions in patients with TM, TI patients experience iron overload because of increased iron absorption. Lately, hepcidin synthesized in the liver is considered as the central regulator in the iron metabolism. In this study prohepcidin, the antecedent of hepcidin hormone levels are determined in patients with TM and TI to determine the role of this hormone in iron accumulation.

Material and Methods: Serum prohepcidin and ferritin levels are determined in 34 patient with TM, 10 patient with TI and 40 control patients without infection/inflammation, hepatitis or liver failure in Adnan Menderes University Medical Faculty Department of Pediatric Hematology and Aydın Atatürk State Hospital. Serum prohepcidin levels were measured using a commercial enzyme-linked immunosorbent assay (DRG International, Inc. Marburg, Germany), ferritin was studied with chemiluminescence method in immulite 2000 machine.

Results: Mean serum ferritin levels in TM, TI and control groups were 2347,97±1724,81 ng/ml (range; 144-8015 ng/ml), 1352,40±918,94 ng/ml (range, 311-3109 ng/ml), 33,35±12,03 ng/ml (range; 20-69,1 ng/ml) respectively. Also serum prohepcidin levels in the same groups were 221,78 ±74, 38 ng/ml (range, 71,14-446,57 ng/ml), 173,31±52,14 ng/ml (range; 100,83-267,69 ng/ml), 218,20±50,37 ng/ml (range; 116,18-330,43 ng/ml) respectively. Only prohepcidin in patients with TI and control group were statistically significantly ($p=0.016$). No correlation was found between prohepcidin and ferritin levels in all groups.

Conclusions: The low levels of prohepcidin in patients with TI may be related to increased gastrointestinal iron absorption and increased erythropoietic activity. Addition of synthetic analogs of hepcidin to the treatment of patients with thalassemia can prevent uncontrolled intestinal iron absorption in these patients.

Key words: Prohepcidin, thalassemia major, thalassemia intermedia, erythropoiesis