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Concept: Duran Canatan, Design: Duran Canatan, Data Collection or Processing: Serpil Delibaş, Gülsüm Yazıcı, Vildan Çiftçi, Analysis or Interpretation: Türker Bilgen, İbrahim Keser, Gülsüm Yazıcı, Vildan Çiftçi, Literature Search: Duran Canatan, Türker Bilgen, Writing: Duran Canatan.

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References

- 1. Harteveld CL, Higgs DR. Alpha-thalassaemia. Orphanet J Rare Dis 2010;5:13.
- Blackwell RQ, Jim RT, Tan TG, Weng MI, Liu CS, Wang CL. Hemoglobin G Waimanalo: alpha-64 Asp leads to Asn. Biochim Biophys Acta 1973;322:27-33.
- Brennan SO, Chan T, Ryken S, Ruskova A. A second case of Hb Fontainebleau [alpha21(B2)Ala-->Pro] in an individual with microcytosis. Hemoglobin 2009;33:258-261.
- 4. Akar N. An updated review of abnormal hemoglobins in the Turkish population. Turk J Hematol 2014;31:97-98.
- Lin M, Wu JR, Yang LY, Chen H, Wang PP, Wang Q, Zheng L. Hb G-Waimanalo: occurrence in combination with alpha-thalassemia-1 Southeast Asian deletion. Blood Cells Mol Dis 2009;42:36-37.
- Tan TG, Jim RT, Blackwell RQ. Hemoglobin G Waimanalo beta thalassemia. Hawaii Med J 1978;37:235–239.
- Lin M, Wang Q, Zheng L, Huang Y, Lin F, Lin CP, Yang LY. Prevalence and molecular characterization of abnormal hemoglobin in eastern Guangdong of southern China. Clin Genet 2012;81:165–171.
- 8. Wajcman H, Blouquit Y, Gombaud-Saintonge G, Riou J, Galacteros F.Hb Fontainebleau [alpha 21(B2)Ala----pro], a new silent mutant hemoglobin. Hemoglobin 1989;13:421-428.
- Turner A, Sasse J, Varadi A. Hb Fontainebleau (HBA2: c.64G>C) in the United Arab Emirates. Hemoglobin 2014;38:216–220.



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Serum Lipids in Turkish Patients with $\beta\text{-Thalassemia}$ Major and $\beta\text{-Thalassemia}$ Minor

Türk β-Talasemi Majör ve β-Talasemi Minör Hastalarının Serum Lipidleri

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To the Editor,

It is well-known that β -thalassemia is associated with changes in plasma lipids and lipoproteins [1,2,3]. To our knowledge, no data are available on lipid profiles in Turkish β -thalassemia major (TM) and β -thalassemia trait (TT) patients together. The aim of this study was to evaluate lipid profiles in two groups of patients with β -TM and β -TT and to compare them with healthy controls. The study included a total of 311 subjects. Group 1 included 131 β -TM patients (mean age: 16.3 \pm 7.58 years). Group 2 included 68 β -TT patients (mean age: 7.25 \pm 4.43 years). Group 3 consisted of 112 age- and sex-matched healthy controls (mean age: 9 \pm 4.7 years). Serum ferritin level was 2487 \pm 1103 (range: 661-5745) ng/mL in Group 1. In comparing the correlation between ferritin and lipid parameters, while a significantly negative relationship was detected between ferritin and high-density lipoprotein cholesterol (HDL-C) (p=0.000, r=-0.602), a

significantly positive relationship was detected between ferritin and triglyceride (TG) levels (p=0.02) in TM patients. Serum lipid profiles of the 3 groups are shown in Table 1.

Previous studies have shown total serum cholesterol, HDL-C, lower low-density lipoprotein cholesterol (LDL-C), and higher TG in β -TM patients compared to healthy controls [1,2,3]. In our study, we found lower serum total cholesterol, lower HDL-C, LDL-C, and higher TG in β -TM patients compared to healthy controls. The pathophysiology of hypocholesterolemia in thalassemia remains obscure, although several mechanisms have been proposed; plasma dilution due to anemia, increased cholesterol requirement associated with erythroid hyperplasia, macrophage system activation with cytokine release, and increased cholesterol uptake by the reticuloendothelial system [4,5]. Previous studies reported different variations in lipid profiles of β -TT patients [6,7]. In our study, we demonstrated

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Table 1. Lipid profiles and their significance in patients with β -thalassemia major, patients with β -thalassemia trait, and controls.

	Group 1	Group 2	Group 3	p-values		
	β-TM (n=131)	β-TT (n=68)	Control (n=112)	Groups 1-2	Groups 1-3	Groups 2-3
T-Chol	118.5±30.6	145.6±27.6	154.3±31.7	0.00	0.00	NS
LDL-C	59.1±27.6	82.5 <u>+</u> 24.9	89.6±26.1	0.00	0.00	NS
HDL-C	34.4±11.2	45.7 <u>+</u> 12.2	45.5±11.1	0.00	0.00	NS
TG	121.8±50.8	82.9±34.6	97.8±52.4	0.00	0.00	NS

T-Chol: Total cholesterol, LDL-C: low-density lipoprotein cholesterol, HDL-C: high-density lipoprotein cholesterol, TG: triglyceride, β -TM: β -thalassemia major, β -TT: β -thalassemia trait, NS: non-significant.

similar lipid profiles in β -TT patients and healthy controls. Based on statistical insignificance, we considered that the effects of lipid profile on the development of atherosclerotic vessel disease were similar in both β -TT patients and the healthy control group. Serum iron and iron stores, expressed as elevated ferritin levels, have been implicated in coronary artery disease. Iron overload depletes the antioxidant and HDL-C levels. Lower HDL-C level is an important risk factor for development of coronary heart diseases [8]. We found significant relationships of serum ferritin levels with TG and HDL-C in β -TM patients. These results indicate that β -TM patients who need life-long red blood cell transfusions should receive chelation therapy not only for iron overload-induced congestive heart failure but also in order to prevent cardiovascular diseases resulting from lipid profile alterations.

In conclusion, lipid profiles of β -TM patients differed from those of β -TT patients and healthy controls. The present study demonstrates that lower levels of HDL-C in β -TM should be a reason for concern for better evaluation of the cardiovascular risk factors in β -TM. In order to reduce the effects of lipid metabolism on cardiovascular disorders, an effective chelating therapy is essential in TM patients.

Keywords: Thalassemia major, Thalassemia minor, Serum lipids

Anahtar Sözcükler: Talasemi majör, Talasemi minör, Serum lipidleri

Authorship Contributions

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References

- Maioli M, Vigna GB, Tonolo G, Brizzi P, Ciccarese M, Donega P, Maioli M, Fellin R. Plasma lipoprotein composition, apolipoprotein (a) concentration and isoforms in β-thalassemia. Atherosclerosis 1997:131;127-133.
- Mansi KM, Aburjai TA. Lipid profile in Jordanian children with β-thalassemia major. Int J Hematol 2008;18:93–98.
- Nasr MR, Abdelmaskoud AM, Abd El-Aal KS, Mabrouk NA, Ismael WM. Plasma lipid profile and lipid peroxidation in beta-thalassemic children. J Clin Lipidol 2008;2:405-409.
- Papanastasiou DA, Siorokou T, Haliotis FA. β-Thalassaemia and factors affecting the metabolism of lipids and lipoproteins. Haematologia (Budap) 1996;27:143-153.
- Hashemieh M, Javadzadeh M, Sihirkavand A, Sheibani K. Lipid profile in minor thalassemic patients: a historical cohort study. Bangladesh Med Res Counc Bull 2011;37:24-27.
- Namazi MR. Minor thalassemia as a protective factor against cerebrovascular accidents. Med Hypotheses 2002;59:361–362.
- Maioli M, Pettinato S, Cherchi GM, Giraudi D, Pacifico A, Pupita G, Tidore MG. Plasma lipids in β-thalassemia minor. Atherosclerosis 1989;75:245-248.
- Brizzi P, Isaja T, D'Agata A, Malaguarnera L, Malaguarnera M, Musumeci S. Oxidized LDL antibodies (OLAB) in patients with β-thalassemia major. J Atheroscler Thromb 2002;9:139-144.



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