Pediatrics & Therapeutics
ISSN: 2161-0665

Arıca et al., Pediat Therapeut 2012, 2:5
DOI: 10.4172/2161-0665.1000130

Research Article Open Access

Serum Lipid Values in Children with Beta Thalassemia Major

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Abstract

Objective: The purpose of the study was to examine the blood lipid profile in children with beta-Thalassemia major, and to determine the factors that affect it.

Material and Method: Files of eighty-five patients between the ages of five and fifteen with beta-Thalassemia major who were receiving regular chelation therapy followed by from paediatric policlinic of our hospital were examined retrospectively. Blood lipid profiles of fifty-five healthy children were taken for use as the control group. A total of 117 children were enrolled and examined in the study.

Findings: Hb and Hct values of the group with Beta-Thalassemia major were significantly lower than the control group ($p<0.005$). Ferritin values in the group with Beta-Thalassemia major were found to be significantly higher than in the control group ($p<0.005$). Cholesterol, HDL-cholesterol, LDL-cholesterol levels were found to be significantly lower in patients with Beta-Thalassemia major than in the control group ($p<0.001$), while the triglyceride level was found to be higher ($p<0.001$).

Result: We determined a positive correlation between triglyceride and serum ferritin levels. It may indicate excessive iron loading, and changes in blood lipid values in patients with Beta-Thalassemia major.

Keywords: Beta-thalassemia major; Lipid profile; Ferritin

Introduction

Thalassemia is an inherited blood disease characterized by inadequate production of haemoglobin globin subunits, ineffective hematopoiesis, and increased hemolysis [1,2]. Thalassemia is seen as the most frequent monogenic disease in the world. Approximately 5% (270 million) of the world's population are thalassemic and abnormal haemoglobin carriers. The frequency has increased especially in Mediterranean countries, the Middle East, the Far East, and also in Europe and America due to migration [3]. Carrying thalassemia is observed very frequently in Cukurova, the Mediterranean coast, and the Aegean and Marmara regions in our country. The frequency of carriers of beta-Thalassemia in the healthy Turkish population is 2.1%. There are approximately 1.4 million carriers and about 4.500 patients in Turkey. Between 1995 and-2000, the Ministry of Health and the National Hemoglobinopathy Council collected screening studies performed by 16 centers in Marmara, Aegean, and the Mediterranean regions, and reported the frequency as 4.3% [4].

Infections such as HBV, HCV, and HIV can be observed through frequent transfusions in children with Beta-Thalassemia major (B-TM) [5,6]. Various endocrine, cardiac, and hepatic diseases may occur depending, on excessive iron-loading [7]. The production of free radicals associated with excessive iron-loading is increased in these patients [8]. In recent years, the relationship between the increase in blood lipid levels and atherosclerotic diseases was shown in the performed researches [9-11]. Blood lipid levels were decreased much less in children with Beta-Thalassemia major [8].

This study is very important in terms of the examination of the blood lipid profile in children with thalassemia major in our country, which is a Mediterranean country.

Material and Methods

The files of eighty-five patients (44 males' and-41 females) between the ages of five and fifteen with beta-Thalassemia major who were receiving regular chelation therapy followed from paediatric policlinic of Mustafa Kemal University, Faculty of Medicine, were examined retrospectively. Sixty-two patients (35 males' and-27 females) whose lipid profile was measured in his or her file were enrolled in the study. Blood lipid profiles of 55 healthy children (30 males' and-25 females) were taken as the control group. A total of 117 children were enrolled in the study and examined. Samples of the blood lipid levels of children were routinely taken on an empty stomach in the morning. HBV, HCV, and HIV results were negative when all of the case files were examined. Chelation therapy has been applied to all children with Beta-Thalassemia major between one and three times per a week (Desferrioxamine, subcutaneous infusion, 2 to 2.5 g). Cholesterol, triglyceride, LDL, and HDL-cholesterol values as the level of blood lipid were measured by using "Olympus System Reagent" kits.

Average values and standard deviations of results were calculated. ANOVA and student t tests were used to compare both groups and $p<0.001$ values were regarded as statistically significant.

Findings

In Table 1, Hb, Hct, and ferritin values of the test and control groups are displayed. Hb and Hct values of the group with Beta-Thalassemia major were significantly lower than those of the control group ($p<0.005$). Ferritin values in the group with Beta-Thalassemia major were found as 2372 ± 1455 in males, and 2286 ± 1614 in females. Ferritin values in the control group were 52 ± 26 in males, and 50 ± 25 in females. Ferritin values in the group with B-TM were found to be significantly higher than those of the control group ($p< 0.005$).

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Received March 12, 2012; **Accepted** July 20, 2012; **Published** July 23, 2012

Citation: Arıca V, Arıca S, Özer C, Çevik M (2012) Serum Lipid Values in Children with Beta Thalassemia Major. Pediat Therapeut 2:130. doi:10.4172/2161-0665.1000130

	patients (n:85)		control (n:55)		p
	male (n:44)	females(n:41)	males(n:30)	females (n:25)	
Hemoglobin (g/dl)	8.8 ± 2.4	9.1 ± 2.6	11.7±0.6	12.0±0.8	<0.005*
Hematocrite (%)	28.0 ± 2.5	29.0 ± 3.7	36±3.2	38±3.6	<0.005*
Ferritin(u/gl)	2286 ± 1614	2372 ± 1455	52±26	50±25	<0.005*

* p< 0.005 statistically significant

Table 1: Hb, Hct and ferritin values in patients with Beta-Thalassemia major.

	cholesterol (mg/dl)	Triglyceride (mg/dl)	HDL-cholesterol (mg/dl)	LDL-cholesterol (mg/dl)	VLDL (mg/dl)
control groups (n:55)	156.1±27.6	71.7±20.4	47.4±7.8	105.4±27.2	16.0±6.9
patients groups (n:85)	118.2±32.0	135.7±44.5	24.0±5.3	62.2±24.6	27.7±8.5

Table 2: Serum lipid levels of patients with Beta-Thalassemia major and the control group.

As seen in Table 2, cholesterol, HDL-cholesterol, and LDL-cholesterol levels in patients with B-TM were found to be significantly lower than those of the control group (p<0.001), while the triglyceride levels were found to be higher (p<0.001). The average total cholesterol values were measured in the group with B-TM and the control group as 118.2 ± 32.0 mg/dl, 156.1 ± 27.6 mg/dl (p<0.001), respectively; the average triglyceride values as 135.7 ± 44.5 mg/dl, 71.7 ± 20.4 mg/dl (p<0.001), respectively; the HDL-cholesterol values as 24.0 ± 5.3 mg/dl, 47.4 ± 7.8 mg/dl (p<0.001), respectively. Average LDL and VLDL values were found as 62.2 ± 24.6 mg/dl, 105.4 ± 27.2 mg/dl (p<0.001); 27.7 ± 8.5 mg/dl, 16.0 ± 6.9 mg/dl (p<0.001), respectively.

Discussion

Serum lipid values vary in patients with Beta-Thalassemia major. In our particular study, we revealed blood lipid levels of children with B-TM in Turkey. There have previously been very few studies in the literature on this subject [12-18].

In performed studies, cholesterol, HDL-cholesterol, and LDL-cholesterol levels were found to be lower than those of healthy individuals [12-16]. The decrease in total cholesterol, HDL-cholesterol, and LDL-cholesterol levels in our study conform to these results. Whereas the triglyceride level does not exhibit significance in some studies performed in patients with B-TM (13-15), it was determined to be high in some studies [12,16-18] The data we have obtained are consistent with the study in direction in that triglyceride levels are higher compared those of healthy individuals. Different results were obtained in studies in terms of explaining the serum lipid changes observed in patients with B-TM. Liver damage [14]. Low activity of hepatic and extrahepatic lipase enzymes, [15] and the quick cleaning of modified HDL and LDL (richer than triglyceride, poor, cholesterol ester) by activated monocytes and macrophages were held responsible. There are many factors for these blood lipid changes in children with B-TM such as excessive iron loading (high ferritin values), liver damage (deterioration of the ratio between AST and ALT) and hormonal disorders [13-16].

Some studies have suggested that low blood cholesterol values may occur as a result of an increase of erythropoiesis in patients with B-TM and increase of LDL uptake by macrophages and histiocytes that exist in reticuloendothelial system (RES) [19,20]. A study demonstrated that total phospholipids and its functions also decrease with the decrease of total cholesterol [21]. In the same study it was shown that the levels of serum lipid multiple unsaturated fatty acids decreased [21]. Those changes appear as a result of excessive iron-loading and liver damage [21]. Serum total cholesterol level has been found to be low, in accordance with the study we performed in patients with B-TM. However, our obtained results are not sufficient to clarify the subject.

We determined a positive correlation between triglyceride and serum ferritin levels in our study. This also suggest us that it may be effective on blood lipid values as a result of excessive iron-loading due to excessive iron-loading, and decrease the efficacy of chelation therapy or its inadequately application. These results may support the hypothesis that both serum iron and serum triglyceride play role in LDL-C oxidation pathogenesis.

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