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Study of Serum Lipid Profile in Beta-Thalassemia Major Patients

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Abstract

Background: Beta-thalassemia is the commonest singlegene disorder in the Indian population. Beta thalassemia major is caused by complete absence of beta globin chain production resulting from reduced synthesis of one or more globin chains which can be caused by different globin gene mutation resulting in ineffective hematopoiesis, increased hemolysis and early onset anemia.

Methods: A cross-sectional; case control study was done on 50 diagnosed Cases of beta-thalassemia major in the age group of 1 year to 18 years receiving regular blood transfusions; not suffering from any ailment or any other disease leading to deranged lipid profile were included. In controls; 50 normal healthy children were included.

Results: Lipid analyses of controls and thalassemic children. It is clear from the results that beta thalassemia major patients had significantly lower total cholesterol (TC), high-density lipoprotein cholesterol (HDL) and low-density lipoprsotein cholesterol (LDL) compared with controls (p<0.001).

Conclusion: Lipid profile in Beta thalassmia patients show significantly low levels of total cholesterol, LDLC and HDL-C.

Keywords: Beta Thalassemia Major, Lipid Profile, Hypocholesterolemia.

Introduction

Beta-thalassemia is the commonest single-gene disorder in the Indian population. Beta thalassemia major is caused by complete absence of beta globin chain production resulting from reduced synthesis of one or more globin chains which can be caused by different globin gene mutation resulting in ineffective hematopoiesis, increased hemolysis and early onset anemia. Ten percent of the total worlds thalassemic are born in India every year. Certain communities in India, like Sindhis, Guajarati's, Punjabis, and Bengalis, are more commonly affected with beta thalassemia, the incidence varying from 1 to 17%. 1-2

It has been estimated that the prevalence of pathological hemoglobinopathies in India is 1.2/1,000 live births, and with approximately 27 million births per year this would suggest the annual birth of 32,400 babies with a serious hemoglobin disorder. Reportedly, there are about 240 million carriers of β -thalassemia worldwide, i.e. 1.5% of world population. A WHO update on betathalassemia in India indicated a similar overall carrier frequency of 3–4%, based on that the current national population would translate to between 35.6 and 47.5 million carriers of the

disorder nationwide. The mainstay of treatment of thalassemia is regular blood (Packed Red Blood Cells) transfusions. The major complications of blood transfusion are those related to transmission of infectious agents or development of iron overload.³⁻⁴

Materials and Methods

A cross-sectional; case control study was done on 50 diagnosed Cases of beta-thalassemia major in the age group of 1 year to 18 years receiving regular blood transfusions; not suffering from any ailment or any other disease leading to deranged lipid profile were included. In controls; 50 normal healthy children were included. After explaining about purpose of study, outcome and explaining that respondent can refuse and withdraw from study at any time in local language an informed consent was taken. All details were recorded in a pre-tested proforma which included personal information, transfusion history, clinical examination and lipid profile of the patients. Lipid profiles were done using auto analyzer. Statistical analyses were performed by using IBM SPSS Statistics 22.

Results

Table 1. Socio-demographic profile

Socio-	Case	Control
demographic		
variable		
Mean age ±SD	7.42 ± 4.16	8.10 ± 4.06
	years	years
Male : Female	31:19	29:21

Out of total 50 cases, 31 were female and 19 were male while out of total 50 controls 29 were female and 21 were male. Mean age of cases was 7.42 ± 4.16 years and of controls was 8.10 ± 4.06 years.

Table 2: Comparison of various parameters of lipid profile in cases and control group

Lipid	Case	Control	p-value
profile			
Total	112.34±26.24	124.25±14.54	< 0.001
Cholesterol			
Triglyceride	111.34±51.24	74.62±16.02	< 0.001
HDL	26.21±9.24	47.42±4.25	< 0.001
LDL	62.24±21.22	76.21±15.31	< 0.001

Lipid analyses of controls and thalassemic children. It is clear from the results that beta thalassemia major patients had significantly lower total cholesterol (TC), high-density lipoprotein cholesterol (HDL) and low-density lipoprsotein cholesterol (LDL) compared with controls (p<0.001).

Discussion

In this study lipid profile in beta thalassemia children were evaluated &compared it with a group of healthy children. In our study it was found that there is significant difference in between the values of cases and controls. It is seen that in beta thalassemia major Patient's serum levels of HDL, LDL and TC is low as compared to controls. In addition of that Triglyceride levels were substantially high. This appears because of many factors. Results of present study correlate well with previous study 5-9

Most studies explain the mechanism of alteration in lipid profile of cases. This alteration is likely due to diminished hepatic biosynthesis as of anemia and iron overload, while a reduced extra hepatic lipolytic activity could account for the rise in circulating TG. Iron-loading and the effects of repeated blood transfusions induce a hepatic acute-phase response, which could determine an LDL-class shift towards protein-rich, denser particles ^{7,8}

Some authors ¹⁰⁻¹² suggested that accelerated erythropoiesis and increased uptake of LDL by macrophages and histiocytes of the reticulo endothelial

system are the main determinants of low plasma cholesterol levels in beta thalassemia major.

Factors such as iron overload (high ferritin level), liver injury, and hormonal disturbances affects lipids pattern among patients with major form of beta-thalassemia, while some researchers observed that the lipid profile in thalassemia major patients is not influenced by age, sex, liver injury, and hemoglobin or ferritin levels. The higher erythroid bone marrow activity with the enhanced cholesterol consumption could be the dominant mechanism implicated in the lipid abnormalities of thalassemia major patients. However other researchers did not find such differences.

Conclusion

Lipid profile in Beta thalassmia patients show significantly low levels of total cholesterol, LDLC and HDL-C.

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