DYSLIPIDEMIA IN BETA THALASSAEMIA MAJOR PATIENTS

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ABSTRACT

BACKGROUND: Beta thalassaemia is the most common frequent hereditary blood disorder throughout the world. Changes in lipid levels have been seen in different types of beta thalassaemia.

OBJECTIVE: The aim of study was to evaluate the lipid profile (serum cholesterol, HDL, LDL, serum triglyceride) in beta-thalassemia major patients and to compare the lipid profile of beta-thalassemia major patients with healthy controls.

MATERIALS AND METHODS: In the present study we included clinically diagnosed 45 patients of beta thalassaemia major. The levels of Cholesterol, Triglyceride, HDLc, LDLc, VLDLc, were measured by using latest techniques in clinical biochemistry laboratory.

RESULTS: Our study revealed that in thalassaemic patients lipid pattern became altered had high level of triglyceride, low level of cholesterol and low HDL-cholesterol levels and conclude that thalassaemic patients are at risk of coronary heart disease.

CONCLUSION: Significant variations were found which confirm that lipid changes occur in B-thalassaemic patients when Total cholesterol, Serum triglycerides, Low density lipoprotein cholesterol, high density lipoprotein cholesterol levels are compared with normal healthy subjects. Multiple factors such as age, iron overload, hormonal changes and liver injury might cause these changes. As this study focus on lipid abnormalities in beta thalassaemia major, this may help clinicians in the management of such patients.

KEYWORDS: Total Cholesterol (TC), Triglyceride (TG), High density lipoprotein (HDL), Low density lipoprotein (LDL), Beta thalassaemia major.

INTRODUCTION

Thalassemia major is a genetic disorder and it results from the inherited defect in the Beta-globin chain synthesis which causes hereditary anemia¹. Thalassemia is a severe public health issue which represents the most common single gene defect. More than 90 million of them carry defective genes leading to thalassemia; Round about 190 million people throughout the world have genetic mutations related with different hemoglobin disorders. As it is common in certain populations, therefore its prevalence is common and in its homozygous state it causes severe anemia.²

The most common monogenic disorder in the world is thalassemia. The gene frequency of beta thalassemia in pakistan is estimated to be 5-8% and it is present in all ethnic groups. It is, therefore estimated that each year more than 5000 children are born with transfusion dependent Beta thalassemia. The average life expectancy of these patients was not more than 10 years in the last decade.³ In various types of hematological disorders likely sickle cell disease, glucose-6-phosphate dehydrogenase (G6PD) deficiency, spheroctosis and aplastic anemia and in different types of beta thalassemia lipid abnormalities have been observed.⁴

Thalassemia major disorder can be prevented. In the Mediterranean this concept has been extensively validated where the new birth rate of thalassemia major has come down to almost zero. However, such type of type of results cannot be gained in any of the developing countries where the disorder poses a significant health problem and fairly common.⁵ There is
reticuloendothelial, severe cardiopulmonary and other major system dysfunction in the patients with beta-thalassemia and scientific evidence have raised the adverse effect of abnormal blood lipid levels, like total cholesterol and other lipids and lipoproteins, on atherosclerotic disease. ^6^7^8^ 

Because of dyslipidemia children with beta thalassemia are at risk of developing premature atherosclerosis. Abnormal lipid profile has been detected in beta thalassemia. ^9^ 

To the best of our knowledge such type of data regarding serum lipids concentration in Pakistani patients with beta thalassemia major are not available. This lack of information encouraged us to determine lipids levels, in comparison to controls and evaluate its significance in Pakistani patients with beta thalassemia major.

**MATERIALS AND METHODS**

Study design: Cross-sectional comparative study

Sample size: 45 major beta-thalassemic patients and 45 controls

Study duration: Six months

Study setting: Children Hospital Lahore from March 2013 to August 2013.

Sampling: Consent was taken from beta-thalassemic patients and control persons. Blood has been taken from both beta-thalassemic patients and control persons. Then it was centrifuged to get serum for cholesterol, HDL, LDL and triglyceroid. Assays were performed in micro lab 300 (semi automated chemistry analyzer Kits of Merck was utilized).

Inclusion Criteria: Diagnosed beta-thalassemia major patients with more than 5-6 transfusions were included.

Exclusion Criteria: Diagnosed cases of cardiac disease, hyperlipidemia, hemochromatosis, Wilsons disease, hypo and hyperthyroidism were excluded.

**METHODOLOGY AND TECHNIQUES:**

Cholesterol - Enzymatic method 3. HDL Cholesterol-Phosphotungstate method 4. LDL Cholesterol - Friedewalds formula 5. VLDL Cholesterol- Friedewalds formula 6. Triglyceride- Enzymatic method 7. Iron-Dipyridyl method. The levels of Cholesterol, Triglyceride, HDLc, LDLc, VLDLc, were measured by using latest techniques in clinical biochemistry laboratory.

**STATISTICAL ANALYSIS**

The data was analyzed by using SPSS software version-16. Z-test was used for statistical significance. P-value < 0.05 was considered statistically significant. Data was presented as mean ± SD. In group-1 beta thalassemic major patients and group-2 healthy control were taken and their results were compared.

**RESULTS**

The results are given in the table 1.1 and 1.2. Regarding age and sex there is no significant difference but when their lipid profile was compared we found significant difference (p-value < 0.05). Mean age of the patients was 23.1± 6.4 and control was 23.7±7.1.

These patients were compared with healthy normal patients on the basis of age, sex, and dietary Conditions and life style.

**Table 1.1 Age and Sex distribution**

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Group-1 (n=45)</th>
<th>Group -2 (n=45)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (year)</td>
<td>23.1 ± 6.4</td>
<td>23.7 ± 7.1</td>
</tr>
<tr>
<td>Sex (Male/Female)</td>
<td>25/20</td>
<td>23 / 22</td>
</tr>
</tbody>
</table>

**Table 1.2 Comparison of lipid profile of beta thalassemia patients with control group**

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Group-1 (n=45)</th>
<th>Group -2 (n=45)</th>
</tr>
</thead>
<tbody>
<tr>
<td>T.cholesterol (mg/dl)</td>
<td>178.08 + 24.24*</td>
<td>263.4 + 32.15</td>
</tr>
<tr>
<td>Triglyceride (mg/dl)</td>
<td>205.18 + 64.92*</td>
<td>122.86 + 24.70</td>
</tr>
<tr>
<td>HDL-C (mg/dl)</td>
<td>39.5 + 6.62</td>
<td>48.76 + 7.06</td>
</tr>
<tr>
<td>LDL-C (mg/dl)</td>
<td>98.10 ± 22.63*</td>
<td>188.29 ± 40.87</td>
</tr>
<tr>
<td>VLDL-C (mg/dl)</td>
<td>41.30 ± 12.98</td>
<td>24.57 ± 4.94</td>
</tr>
<tr>
<td>Hb (g/dl)</td>
<td>9.5 ± 2.8*</td>
<td>11.4 ± 0.2</td>
</tr>
<tr>
<td>Serum iron (ug/dl)</td>
<td>165.55 ± 14.30*</td>
<td>112.14 ± 15.28</td>
</tr>
</tbody>
</table>

*Significant difference compared with controls (p < 0.05)
DISCUSSION
Changes in lipid pattern has been consistently reported in B-thalassaemia, but its pathogenesis is not exactly clear\textsuperscript{9,10}. In the present study, we found elevated triglyceride, low total cholesterol, low LDL cholesterol and low HDL-cholesterol in beta thalassaemia major patients in comparison with control subjects. Our results agree with previous findings in patients with beta thalassaemia major whose lipid profile altered\textsuperscript{11,12}. This alteration is probably due to a reduced extrahepatic lipolytic activity could account for the rise in circulating TG and decreased hepatic synthesis because of anemia and iron overload\textsuperscript{13}. In 1991 Goldfarb and colleagues suggested that accelerated erythropoiesis, liver damage and an increased uptake of low-density lipoprotein (LDL) by macrophages and histiocytes of the reticuloendothelial system are the main determinants of low plasma cholesterol in B-thalassaemia\textsuperscript{14,15,16}. In this study we found that serum TG was significantly elevated in comparison with controls. In 2007 a study by Amendola and colleagues suggested that the higher bone marrow activity with enhanced cholesterol consumption could be the cause of lipid abnormality in thalassemia\textsuperscript{17}.

Bersot et al\textsuperscript{18,19,20} suggested that risk for coronary heart disease is caused by low HDL cholesterol. In our study we found that HDL cholesterol values were low and we compared these results with other studies which shows similarity and studies also suggest that risk for myocardial infarction is high when HDL cholesterol is low. This gives us the importance of Total-to-HDL cholesterol ratio for the evaluation of blood lipids and the prevention of atherosclerotic disease. We could suggest that thalassaemic patients are at much higher coronary risk than their matched controls, because of the low HDL cholesterol production, even if they are within normal values of total cholesterol.

CONCLUSION
Our study revealed that in thalassaemic patients lipid pattern became altered with high level of triglyceride, low level of cholesterol and low HDL-cholesterol levels. We conclude that thalassemic patients are likely at risk of coronary heart disease due to these lipid abnormalities. Therefore awareness of lipid abnormality is helpful for proper overall management of these patients.

REFERENCES

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