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RESEARCH ARTICLE

Estimation of Alpha-L-Fucose and Vitamin D_3 Levels in Beta Thalassemia Patients in Al-Najaf Province

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Abstract

The present study deals with the prominent inherited disease that is thalassemia. B-thalassemia is an extreme hereditary blood disorder resulted after a change in the gene encoding for the hemoglobin ß chains. Hemoglobin is made up of a genetic defect in the synthesis of a globin chain, or more, which results in a reduced rate of these chains. The creation of atypical hemoglobin molecules is a result of the reduction in the synthesis of one chain of the globin, and thus, leads to the most specific symptom of the thalassemia; anemia. This study was conducted with 40 subjects, whose age ranges from 4-20 years, divided among (males 30 and females10), who has visited the Thalassemia Center at Al-Zahraa Teaching Hospital in Najaf province, Iraq, for managing the disease, and 40 voluntary healthy as a control group. The study has been done between September and February 2019. This study was performed to evaluate the levels of serum of Vitamin D_3 and L-Fucose in major and intermediate beta-thalassemia patients. It also estimates protein-bound fucose (PBF), hemoglobin (Hb), hematocrit (HCT), liver function tests, calcium, TIBC and Ferritin. The results showed a significant increase in the levels of alpha -L- fucose and protein-bound fucose in thalassemia patients as compared with control. The results showed a very important reduction ($P<0.001$) in Vitamin D_3,Hb, HCT, TIBC and calcium in thalassemia patients in both males and females in comparison with the healthy control, except ferritin, AST,and ALT which is increased significantly($p<0.001$) in those patients in comparison with control.

Keywords: *Thalassemia, Vitamin D, L-Fucose.*

Introduction

The word “thalassemia” has been derived of the Greek term “Thalassa” (which means sea) and “Haema” (which means blood) and mentions the disorders related to imperfect creation of α- or ß-globin subunits of hemoglobin (Hb) A (α2; ß2), considered as pathologic alleles of one or more globin genes which are positioned on 11 (ß) and 16 (α) chromosomes. However, nearly 200 point alterations or removals which damage translation, processing, or transcription of α- or ß-globin mRNA were acknowledged.

The medical indicators are varied, ranging from the absence of signs to deep deadly anemias inutero, or, where no treatment, in early childhood [1]. Vitamin D (VD) is serious for skeleton mineralization as well as for calcium (Ca) homeostasis, particularly within quick growing periods, specifically pubertal and infantile growing periods.

Osteomalacia (a mineralization deficiency of bone tissues) and rickets (a mineralization deficiency at the bone tissues and epiphyseal growth plates) both are results of Vitamin D Deficiency (VDD) [2]. L-Fucose is a regular essential of glycoproteins and is a monosaccharide which exists at low concentrations in serum [3]. Fucose is obviously could be found in D- and L-form, whereas L-fucose is found in mammalian tissues and fluids. There are two different forms L-fucose; α-LFucose (29.5%) and ß-L-fucose (70.5%) [4].

In normal metabolism process, one of only a few L-sugars, which are regularly used by human beings, is L-Fucose (6-deoxy-L-galactose) which is a 6-carbon deoxyhexose. The only enantiomer of fucose recognized in mammalian glycoproteins is the L-enantiomer.

In these glycoproteins, the terminal sites at the non-reducing ends of oligosaccharide chains are normally occupied by sialic acid and L-fucose. Fucose could also be found in

many membrane-associated glycolipids. With a particular carrying system, L-fucose is usually collected in the eukaryotic cells. In some cell-surface oligosaccharide ligands, which intermediate between adhesion-signaling and cell-recognition passageways, L-fucose could be found at the pre-terminal or terminal sites.

These contain typical actions, for example; early recognition of blood group and embryologic development, and pathologic procedures which include neoplastic progression, recognition of infectious disease, and inflammation [5]. Fucose can be found in blood cell antigens, which are involved in determining blood type. It is distributed in macrophages [6].

Materials and Methodology

This study is case-control carried out in the Center of Thalassemia at Al-Zahra'a Teaching Hospital in Najaf province, Iraq, between September and February 2019. A 40 ß-thalassemia major and intermedia patients (males 30 and females10) were enrolled in this study; the study also involves 40 healthy controls. Both patients and control were age and sex matched and ranged from 4 to 20 years. From beta-thalassemia patients and healthy control, a 5ml of blood has been withdrawn vein puncture.

Estimation of Total Fucose TF

Estimation of TF in accordance with Dische and Sheetels method: This procedure depending on a direct reaction response of concentrated H2SO4 with serum contents. The colored product is formed after mixing the reactants with cysteine, and the color product is measured at (390nm and 430nm). The differences in absorbance were proportioned to □-L-fucose compound in the solution.

Estimation of Serum Ferritin

The levels of serum ferritin are estimated according to biomerieux kit.

Estimation of Serum Vitamin D

The serum vitamin D estimated by ELISA kit from (Calbiotech, USA).

Statistical Analysis

Software of Statistical Package for Social Sciences (SPSS) program version 25 has been used to analyze the data. A p-value of (<0.05) has been taken as significant and (<0.001) was considered very significant.

Result and Discussion

This case-control study included 40 thalassemia patients conducted at thalassemia center in Najaf province, Iraq, and an equal number as a control group. The current study revealed a significant increase in the levels of alpha -L- fucose and protein-bound fucose in thalassemia patients as compared with control, which indicates to clinical severity and increases complications of the disease (table 1). The mean level of serum for 25-OH Vit D was considerably lower than in controls at (P<0.001) in our thalassemic patients (Table 1). This can be explained by repeated blood transfusions with resulting in iron overload.

In the current study, plasma 25-hydroxy vitamin D levels are found to be pointedly low in patients compared with the controls, the result agrees with [7]. The decreased in the level of vitamin D in thalassemia patients came from the deposition of the iron in the patients' skin and liver with thalassemia major. Thus, this could disturb production and hydroxylation of vitamin D. Therefore, we may see that there is a vitamin D lack in most patients with thalassemia major [8].

Comparing with the control group, our results agreed with) [9, 10, 11] that there is a noticeable increase in the levels of alkaline phosphatase and decrease at (P<0.001) in calcium level in thalassemia patients (Table 1). Calcium is an inorganic element that has a significant role in numerous physiological processes in the body, such as nerve impulse transmission, blood clots, heart and smooth muscles, and the contraction of striated muscles.

In the current study, the levels of calcium of patients with thalassemia were considerably less than that of healthy controls (Table 1). This result is consistent with [12] Similar results were also obtained by [11] in Pakistan, [13] and [9] in India. This study found serum ferritin levels in the patient group to be significantly higher at (P<0.001) than those in the control, while TIBC was significantly lower in patients compared to healthy participants (Table 1). The result of

correlation shows a significant negative correlation between L-fucose and vitamin D3 level in beta-thalassemia patients (Figure 1).

Table 1: Total Fucose and Protein – Bound Fucose, Vitamin D3, Ferritin, Calcium, Total iron binding capacity, Biochemical parameters in B-Thalassemia Patients and Control

Parameters	Group		P-value
	Patient N=40	Control N=40	
	Mean ± S.D	Mean ± S.D	
Fucose mg/dl	19.68 ± 6.72	15.32 ± 3.83	<0.001
PBF mg/dl	13.007 ±3.27	10.79± 3.98	<0.001
Vit.D_3ng/ml	16.24±6. 9	28.41± 6.54	<0.001
Calcium mg/dl	7.27±1.74	9.76 ± 0.49	<0.001
Ferritin ng/ml	2700.21±2218.45	81.68±16.94	<0.001
TIBC	48.06 ± 16.3	52.25 ±18.3	<0.001
ALP U/L	243.67 ±92.25	112.36 ± 149.76	<0.001
AST U/L	41.72 ± 15.34	22.77±8.27	<0.001
ALT U/L	39.10 ±18.96	20.09± 9.32	<0.001

Figure (1): Negative correlation between L-fucose and vitamin D3 level in beta-thalassemia patients.

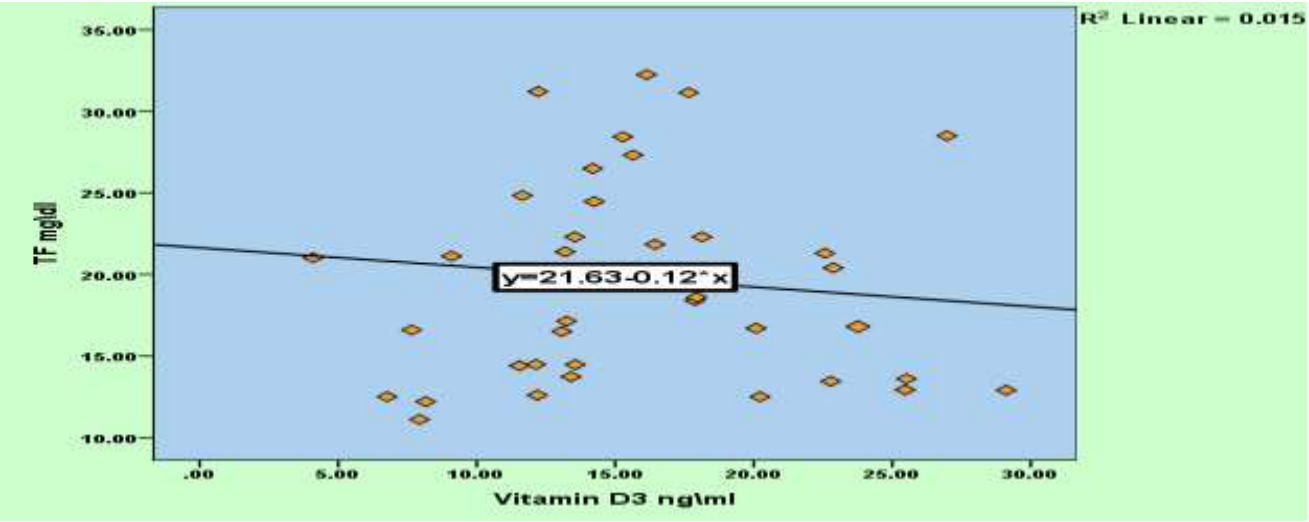

Figure 1: correlation between total fucose and Vitamin D3 for Beta-thalassemia group

Conclusion

The study showed a significant increase in the levels of alpha -L- fucose and protein-bound fucose in thalassemia patients as compared with control, which indicates to clinical severity and increases complications of the disease. The mean level of serum for 25-OH Vit D was considerably lower than in controls at (P<0.001) in our thalassemic patients.

However, plasma 25-hydroxy vitamin D levels are found to be pointedly low in patients compared with the controls. The decreased in the level of vitamin D in thalassemia patients came from the deposition of the iron in the patients' skin and liver with thalassemia major and, therefore, this could disturb production and hydroxylation of vitamin D. There is a noticeable increase in the levels of alkaline phosphatase and decrease at (P<0.001) in calcium level in thalassemia patients. The levels of calcium of patients with thalassemia were considerably less than that of healthy controls.

It has been found serum ferritin levels in the patient group to be significantly higher at (P<0.001) than those in the control, while TIBC was significantly lower in patients compared to healthy participants. The result of correlation showed a significant negative correlation between L-fucose and vitamin D3 level in beta-thalassemia patients.

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