Physiological study for some blood parameters in patients with major β-Thalassemia in Sulaymaniyah, Kurdistan, Iraq.

Assist lecturer Firas F. Rija

College of Nursing , Tikrit University

Abstract

β-thalassemia is an inherited hemolytic disorder caused by partial or complete deficiency of α or β- globin chain synthesis. The disease leads to serious health problems unless treated by regular blood transfusion and iron chelating therapy. The present study carried out on (90) males and females their age from (12-30) years with β - thalassemia during spring of 2014 , there were (70) of them infected with β – thalassemia (major) , who registered at Center of thalassemia and inherited blood disorders, Shaheed Hemin teaching hospital, Sulaymaniyah, Kurdistan, Iraq , while (20) healthy used as a control group. The results present study showed that a significant decrease ($p < 0.05$) in number of red blood cells and the concentration of heamoglobin and packed cell volume (PCV) of infected patients with β- thalassemia major compared with control group , while the results showed increase in the total number of white blood cells in infected objectives with the disease compared with the control group and this increase was a significant increase ($P < 0.05$), as the current research patients had blood group (O) were the most susceptible to the infection with β-thalassemia compared with the other blood groups , as the infected objectives who carry the factor rhesus positive (Rh+) were (% 76) higher than the patients with rhesus factor negative (Rh-) (24 %).

Introduction

Thalassemia is the most important diseases of hemolytic anemia, and it is a Greek word originally that means Mediterranean anemia ,this disease was known and resounded in this region significantly so-called anemia Mediterranean(1,2)and was diagnosed and known their kinds by scientists Lee and Cooley in 1925,so it was called as Cooley's anemia in the United States (3).

Thalassemia is one of an inherited anemias caused by partial or complete deficiency of α or β-globin chain synthesis (4).

Abnormalities in the synthesis and structure of the globin chains that form tetramers of hemoglobin (α2,β2) lead to this disease (5). The types of thalassemia (alpha and beta) which undergo all genes to amutations are the species most dangerous to a pantient's life (6),alpha- thalassemia major causes hydrops fetalis of the embryo and his death before he was born, either β- thalassemia major causes severe anemia, life threatening and the patients need to blood transfusion periodicity each(3-4)weeks according to age

and lack degree of hemoglobin (7). as a result of the inability of the body to form normal red blood cells, but the red blood cells are abnormal and small-sized and full of inculsion bodies that make up the result of the accumulation of alpha and beta chains inside the corpuscular making hemoglobin insoluble and this slushy abnormal condition which the hemoglobin is dissolved in corpuscular(8,9)or a result to continuation production of alpha chains and the lack or absence of the production of beta chains leading to accumulation the alpha chains in corpuscular causing broken the plasma membrane and damage the corpuscular in bone morraw(10,11). and when abnormal corpuscular exit to blood cycle would remove by the spleen, causing severe anemia(12) .

The constant blood transfer causes a condition called iron accumulation resulting from the deposition of iron in the body of the patients gradually leading to derangement of the functions of the cells and thus her death(13,14) and affects many organs of the body, such heart, liver, lungs، kidney and the endocrine glands causing failure(15), beta-thalassemia major may detect during the first months six from the child's age, and the disease leads to a number of complications if not treated as yellowing of the skin (jaundice) and enlargement of the liver and spleen and growth retardation in addition to malformations in the bones of the face and diabetic and disease of the heart muscle(10,16). Beta-thalassemia major are diagnosed by examining the haemoglobin etectrophoresis movement ,where a high amount of fetal hemoglobin due to loss of beta chains leads to increased production of gamma chains which combine with alpha chian to form the fetal hemoglobin which is very familiarity to oxygen and prevents the liberation to the tissue(17,18) .the aim of this study to detection some physiological blood parameter ,blood group and factor rhesus related with β-thalasemia.

Materials and Methods

-Patients

The present study included exame (70) patients (male and female) who were ten years of age and over with β-thalassemia and who are reviewing periodically the Center for thalassemia and inherited blood disorders, Shaheed Hemin teaching hospital, Sulaymaniyah, Kurdistan, Iraq. and then the results that obtained compared with the control group that included (20) healthy male and female .

-Questionnaire

Information was collected by preparing special Questionnaire recorded the necessary information and special of the research samples have included the age / blood group/rhesus factor.

-Take of blood samples.

Blood samples were collected by using medical syringes and then

the samples placed in a tubes container the material anti-coagulation (EDTA), to do blood tests included in this study .

Study the physiology blood parameters.

-Count the number of red blood cells (RBCs) .

Used absorbent with Bead red ,the blood was pulled to the mark (0.5) and then pull the Hyem's solution to the mark (101), Shake the mixture lightly, then put a drop of on a slide count and left the slide to stabilize the cells and then count cells under (40 X) and then extracted the number of red blood cells by applying the following equation(19):-

Red blood cell count/mm3 = N× 80/400 × 200

N: number of red blood cells calculated .

Estimate the concentration of hemoglobin. -

Use solution's Draken who put (5 ml) in the tube, then Added (0.02) ml of blood sample to the tube and shaked the tube well, and left for (10) minutes and then placed in a hemoglobinmeter with wavelength (450)nm (20).

-Measurement the packed cells volume(PCV).

Put amount of blood in the capillary tube to three quarters and then fill one end with the mud artificial ,then tube placed in a microcentrifuge and the exact role of the fast (10000) cycle / minute for (5) minutes and then read a percentage of the packed cells volume(PCV) by using special standard ruler (21).

-Count the total number of white blood cells(WBCs) .

The blood was hauled by the special pipette after the blood which has the white bead to the sign (0.5) .Then, with the same pipette diluted solution was hauled to the sign (11)and shaked the pipette slightly and put a drop of the diluted blood on the count slide after putting a cover on it ,the slide was left in order to make the cells be stable ,then it was examined under small force (10x).In the four big squares (the 64 small squares) ,the white blood cells were counted in mm3 according to the following equation (20):-

Total count of white blood cells in mm3 = Nx4/10 x20

N: number of counted white blood cells.

-The Statistical analysis .

Analysis the results were by using F-test with a significant level ($p<0.05$)for showing the results significant (22).

Results

Effect β- thalassemia major on (RBCs count) , (Hb) , packed cell volume(PCV).The results showed a significant decrease ($p<0.05$) in the (RBCs) and (Hb) and packed cell volume in patients with β-thalassemia major (3.75±0.36 , 6.72±1.7, 20.16±0.56) respectively compared with the control group(5.1±0.53 ,

12.9±0.89 , 38.7±0.76) respectively (Table 1).

Effect β- thalassemia major on the WBCs total .Table(1)showed that a significant increase ($p<0.05$) on the WBCs total in blood of patients with β-thalassemia(13.9±0.97) compared with the control group(8.55±0.58).

The correlation between the blood group and Rhesus factor(Rh) and β-thalassemia major(figure 1). The results of present study revealed that the ratio of patient with β-thalassemia major from blood group O (%43)more infection with the disease ,then patients with blood group A (%36),then patients with blood group AB(%15) and last patients with blood group B(%6). as the infected patients who carry the factor rhesus positive (Rh+) were (%76) higher than the patients with (Rh-) (24 %) (figure 2).

Discussion

Effect β- thalassemia major on number of (RBCs) ,(Hb) and (PCV). The results showed a significant decrease in the RBCs in patients when compare with the control group (Table 1) . The cause returns to the genetic mutations that occur in the genes responsible for the synthesis of protein chains in hemoglobin which lead to disorder in the biosynthesis of the chains globin and thus loss the imbalance in the manufacture of hemoglobin (23) , and this pathology is characterized by decreased Hb production and red blood cell (RBC) survival , resulting from the excess of unaffected globin chain and causing marked RBC damage and severe hemolysis associated with ineffective erythropoiesis (IE) and extramedullary hemolysis (24).

The result of this study are consistent with the results of several studies(3,25). Either decrease in hemoglobin concentration was due to a genetic defect in synthesis globin chains leading to the lack or low production globin chains(α, β)in hemoglobin(26)and this leads to the production of a small number of red blood cells that are small in size and hypochromic (11,13) as well as broken a large number of red blood cells due to a significant decrease in hemoglobin concentration(6). (and this result agreed with the results of many studies(3,25).

Table(1)showed that a significant increase ($p<0.05$) in the WBCs total in blood of patient with β-thalassemia major and showed a significant decrease (, $p<0.05$) compared with the control group.

The increase of the total WBCs in the blood of children with β-thalassemia major may be due to break high percentage of red blood cells inside and outside bone marrow which stimulates secretion erythropoietin hormone from the kidney which stimulates the bone marrow to increase formation of different blood cells included white blood cells (27,16)These results agreed with results of some studies(12 ,25).

Either the relationship between blood groups and factor Rhesus and repeat infection with β-thalassemia Figure (1 and 2) is observed that the blood group (A) and patients with factor Rhesus

positive(Rh+) were more compared with the other blood groups may be due to the small sample or because of varying proportions of these groups in community.

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Table(1):- Effect β- thalassemia major on number of the red blood cells , concentration of hemoglobin and packed cell volume(PCV)

Samples	RBCs Count million / mm^3	Hb mg/dl	PCV %	Total WBCs count cell/mm^3
Control Group	5.1±0.53	12.9±0.89	38.7±0.76	8.55±0.58
Patients	3.75* ±0.36	6.72* ±1.7	20.16* ±0.56	13.9* ±0.97

represents a significant difference at the control group. *

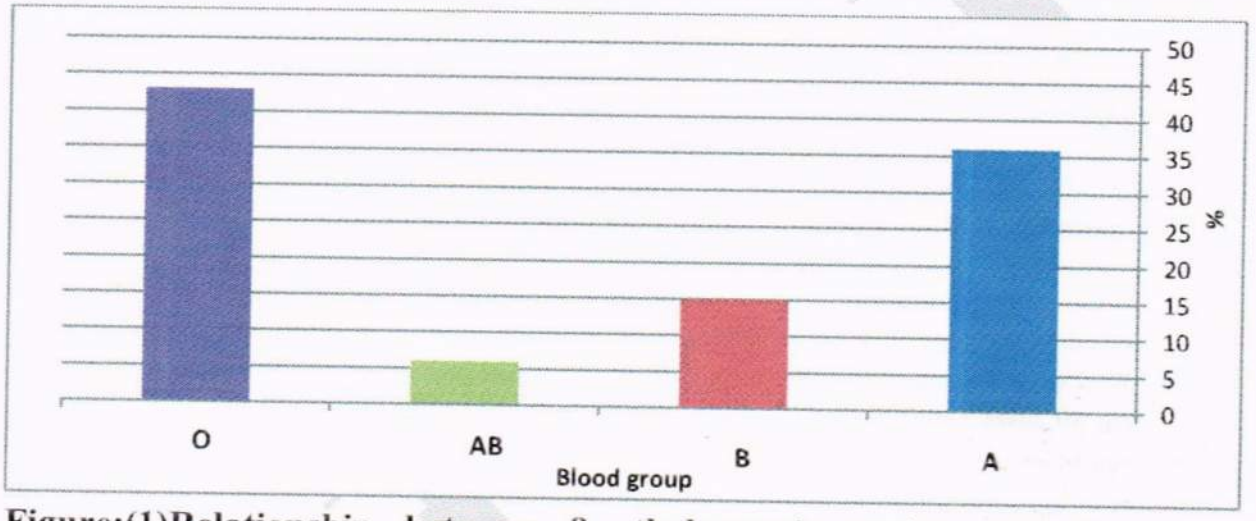

Figure:(1)Relationship between β- thalassemia major and the Blood groups .

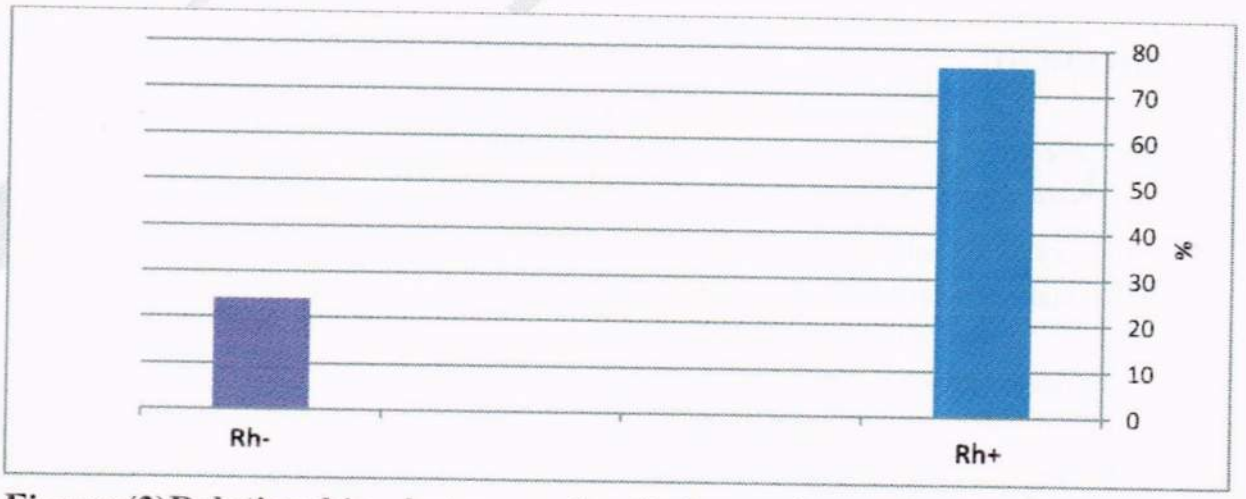

Figure:(2)Relationship between β- thalassemia major and the Rhuses factor(Rh).