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Study of some blood physiological parameters for patients with Beta-thalassemia in AL- Muthanna province – Iraq

Mohammed QasimWaheeb
AL- Muthanna University –Iraq
College of Science

Abstract

The research includes the study of some in changes that occurs in some blood physiological parameters in children infected with the genetic anemia (Thalassemia) in AL- Muthanna province, the study included the examination of 150 children (male and female) infected with their review of the Thalassemia Center Children's Hospital in AL-Muthanna, also used 150 samples from healthy children for period from November 2013 to May 2014 for comparison ranging in age from 2-12 years old. Results of the study have shown high-significant of white blood cells count (WBCs) ($P < 0.05$) as numbered in male and female patients is $(24.5\pm1.1)x10^3$ \mm3 and $(25.5\pm5.1)x10^3$\mm3 cells respectively as compared with healthy children as numbered in males and females is (8.1 ± 1.7) $x10^3$ \mm3and (8.03 ± 1.7) $x10^3$ \mm3 cellsrespectively. A decrease-significant ($P<0.05$) in the level of hemoglobin concentration (Hb) as reached in male and female patients is (7.4±1.15) g/dland (7.6±1.12) g/dl respectively, while in the healthy(male and female) children is (12.3±1.15) g/dl and (12.1± 1.14) g/dl respectively. A decrease-significant ($P < 0.05$)showed in packed cell volume (PCV)as reached in male and female patients is (23.4±2.7) L/ L%, (25.04±3.6) L/ L% respectively, compared with healthy children males and females% L/ L (38.1±2.4) L/ L%, (37.5±2.6) L/ L% respectively. Decrease-significant ($P<0.05$)showed in (RBC) count where its count in males and females patients is (2.9±0.82)x106\mm3 and (2.7±0.6)x106\mm3 cells respectively in comparison with healthy group where its count in males and female is (4.1±0.8)x106\mm3 and (3.7±0.8)x106\mm3 cells respectively.

Key words : Thalassemia , physiological parameters ,anemia.

Introduction:

The thalassemia also called genetic anemia as well as anemia (Mediterranean hereditary disease) because of genetic abnormalities in the formation of natural

polypeptidechains of globin which the molecule hemoglobin, which leads to reduction in the construction of one chains globin or more thereby be partial hemoglobin abnormal and thereby become red blood cells are small size (Microcytosis) and few pigment hemoglobin (Hypochromic) (11). Resulting from the occurrence of Point mutation of a structural gene of Globin responsible for the formation of chains of amino acids, this leads to a reduction or complete absence in the production of hemoglobin chains (12).

Thalassemia can be divided depending on the chains into two main types Alpha-Thalassemia resulting from the mutation of a series of alpha, and Beta-Thalassemia resulting from the mutation of chain beta (12). There are two types of mutations and that gets to Gene Beta B-Gene the first caused a complete absence of production globin chains, therefore called [Thalassemia B^o] and the second cause reduction or lack of production of beta chains called [Thalassemia B+] (15).

Proper human hemoglobin consists of three main parts [A] adult hemoglobin percent (96-98)%, [A2] minor adult Hemoglobin percent (2-3)% and fetal hemoglobin [F] percent (1-2)% (21). The hemoglobin adults [A] consists of four parts protein called poly peptide chain is two alpha chains and two Beta chains $\alpha 2\beta 2$, while the pattern [A2] consists of two alpha chains and two delta chains $2\delta 2\alpha$, while the pattern [F] has two alpha chains two Gama chains $2\ \gamma 2\ \alpha$ (3) (22). When the occurrence of deficiency or absence of one or alpha-beta chains,when the person infected by thalassemia (13).

Thalassemia was descripted for the first time in 1889 by Vondaksh, while the major description of Beta-Thalassemia by pediatrician Thomas Cooley in 1925, in addition Lee, while known the disease Beta-thalassemia minor in 1940 by Wintrob (14), and thalassemia compound word in Greek, the word (Thalassa) means Sea and the word (emia) means anemia Thalassemia means sea anemia (11).

Diseasebeta (major and minor) are widely spread in the population of the Mediterranean basin, including (Greece, Turkey, Italy, as well as the spread in the population of the Middle East, including (Iraq, Palestine, Iran, Jordan), while noting the alpha-thalassemia in the Indian subcontinent them (India, Pakistan) ,as well as the disease extends to Southeast Asia (China) (16).

Thalassemia patients suffering from severe anemia or moderate Anemia or mild anemia and appear red blood cells unusual in different size with a low percentage of hemoglobin to less than 8g /dl, Some the people living to twenty or thirty years of age suffering from severe anemia, loss of appetite, weight loss and sometimes death as a result of the failure of many of the

members of the body, including the liver and spleen due to the accumulation of iron overload (19). This leads to the emergence in the abdomen and swell due to an enlarged liver and spleen (17). In order to prevent the accumulation of iron in the body is given a drug called Desferrioxamine or Desferral to get rid of excess iron in the body where it combines with iron and poses outside the body (18). Incidence of Thalassemiaincreased by inbreeding, as the disease can be transmitted through marriage if strangers were carrying the genetic trait that causes the disease (20).

1-1:The aim of study

The interest including childhood of the most important criteria by which to measure the progress of society and its development, so we have this study to find out the criteria physiological blood for children with thalassemia, including white blood count (WBCs Count) and Hgb concentration of hemoglobin and hematocrit ratio P.C.V. Or Hct and RBCs.

Materials and Methods:

2-1: Materials:-

2-1-1: Devices used:

Table (1) the hardwaredevices,equipment used in the process of analysis for the study

sequence	manufacturer	device name	origin
1-	Sysmix KX – 21	Complete Blood Picture	Japan
2-	Cell-Dyn Ruby-3200	Complete Blood Picture	French

2-2:Methods:

2-2-1 :Method of sample collection:

Collected 150 samples from the blood of patients(75 male, 75 female) with thalassemia type beta and 150 blood samples from healthy children (75 male,75 female) and were withdrawn 2 ml of venous blood and placed in test tubes container material supplied by anti-coagulant EDTA in order to calculate the number of WBCs and the percentage of hemoglobin Hgb and the percentage of Hct or P · C · Vand RBCs.

2-2-1-1The assay of work:

WBCs, hemoglobin concentration Hb, packed cell volume Hct and red blood cells been estimated by placing a sample of venous blood in a complete blood count Complete Blood Picture where the pulls 50µl of blood and is then recorded all blood tests mentioned above directly by the device CBC (32).

2-3 Stastical analysis:

All values were expressed as means ± SE. The data were analyzed by using of computer SPSS program and taking p <0.05 as the lowest limit of significant. Student's t - test was used to examine the differences between different groups. Both t test and ANOVA test were applied to determine the differences between group and another and among all group and within group (33).

3-Results:

Tables (2): Mean of some blood physiological parameters for patient Beta thalassemia and healthy children (male& female) in AL-Muthanna province.

Gender	WBCs x 10^3 Cell/ mm^3	Hb g/dL	P·C·V·orHct L/ % L	RBCx $x10^6$\mm^3
Male patients N=75	24.5±1.1	7.4±1.15	23.4±2.7	2.9± 0.82
Female patients N=75	±5.125.5	7.6± 1.2	25.04±3.6	2.7± 0.6
Healthy male N=75	8.1± 1.7	12.3± 1.15	38.1±2.4	4.1± 0.8
Healthy female N=75	8.03±1.7	12.1± 1.14	37.5±2.6	3.7± 0.8

Values are mean ± SE, p <0.05 .

3-1:White Blood Cells:

The results of the current study showed highly- significant difference (P<0.05) in the number of white blood cells in patients with beta- thalassemia males and females, reaching the average number of white blood cells in each of the male

patients (24.5±1.1) x10³ Cell/ mm³white blood and female (25.5±5.1)x10³ Cell/mm³ white blood cell, as compared with the number of white blood cells in healthy which amounted to (8.1±1.7)x10³ Cell/mm³ in the white blood cell in males and (8.03±1.7)x10³ Cell/mm³ white blood cell in females as in the table (2).

2-3:HemoglobinConcentration

The results of the present study showed, low significant difference (P<0.05) in the concentration of hemoglobin in patients with beta-thalassemia male and female, as the concentration of hemoglobin rate in each of the male patients was (7.4±1.15) g\ dl and females (7.6±1.2) g\ dl as compared with the rate of hemoglobin concentration in healthy people (12.3±1.15) g\ dl in males and (12.1±1.14) g\ dl in females (table 2).

3-3:Packed Cell Volume:(P·C·V.)

The results of the current study showed a significant decrease (P <0.05) in the percentage of the packed cell volume in each of the male patients (23.4 ± 2.7) L/L %, and females patients (25.04 ±3.6) L/ L%, as compared with the healthy males children (38.1 ± 2.4) L/ L % and females (37.5 ± 2.6) L/ L % (table 2).

3-4:Red Blood Corpuscular:

The results of the present study, presence significant difference (P<0.05) in the number of red blood cells in patients with beta- thalassemia males and females, reaching the average number of red blood cells in each of the males patients (2.9 ± 0.8) x 10^6\ mm^3erythrocyte blood and females (2.7±0.6) x10^6 \ mm^3 erythrocyte blood as compared with the healthy which amounted to (4.1±0.8)x10^6\mm^3 erythrocytein males and (3.7.1±0.8)x10^6\ mm^3erythrocyte blood in females (table 2).

4-Discussion

4-1:White Blood Cells

The results of the current study, showed a highly number of white blood cells in children with beta-thalassemia males and females as compared with healthy peopletable (2) and our

results showed that white blood cells increased (29) because of higher crashing to red blood cells inside and outside the bone marrow that causes severe decrease in the partial pressure of oxygen , this is catalyst the production of the hormone Erythropoietin in kidney and this hormone stimulates the bone marrow to increase blood by configure the conversion of non-differentiated cells into differentiated cells. Bone marrow have the ability to configure the types of blood cells, including white blood cells as well as the cells begin to liver cells and some cells spleen configured centers generation a phenomenon outside the blood-forming bone marrow blood cells (24).

The other reason for the rise in the number of white blood cells due to the function of these blood cells, which are the first defensive line in the body against pathogenic bacteria, viruses and other foreign objects and as a result of frequent blood transfusions factors that occur each (14-30) days on what accompanies this process of transition pathogens with blood transfused to patients with thalassemia which injuries that have existed continuously Infection reason that stimulates white blood cells against the presence of factors in the transfused blood, leading to a height (26) (27).

4-2:HemoglobinConcentration:

Evidenced by the results of current study, as shown in the table(2), a reduction in the level of hemoglobin in patients with beta-thalassemia in males and as compared with healthy These results identical with those obtained by the researcher (14).

The reason for the low level of hemoglobin in patients with beta-thalassemia could be due to the turmoil in the manufacture of protein chains(28), and decrease manufacturing alpha chains or beta impedes units hemoglobin manufacturing as the hemoglobin consists of Hem and Protein Globin so the red blood cells are few in number and small size and low hemoglobin Hb (14).

4-3:Packed Cell Volume:(P·C·V·)

Also shows in the current study decline in packed cell volume (table 2) may be because of the rate of P · C · V · depends mainly on the number, size and shape of red blood cells and the extent relieve of blood and his viscous (30), in case of injury Thalassemia

affected red blood cells in terms of the number and characteristics of morphological and volumetric during its formative stages within the bone marrow, As tend to the fact that small in size configured as well as the accumulation of peptide chains is associated beside the membrane and the formation of objects contained(31), this makes them prone to break down by phagocytic cells(Macrophage), which are abundant in the fabric of the bone marrow, which can diagnose and characterize abnormal cells and phagocytic leading to destroy a group of red blood cells through the stages of development (25), but when they enter the circulatory system, they may become vulnerable to self-destroy the inside of blood vessels or may shatter within the spleen through effective phagocytic cell (Macrophage) in the fabric of the spleen and specialized to rid the blood of deformed cells and older and foreign, and on the other hand, the small size of these cells does not make them occupy the same volume occupied by the normal cells (23).

4-4 Red Blood Corpuscular:

Evidenced by the current study, as shown in the table (2) a decreases significant in the red blood count in patients with beta-thalassemia males and females as compared with healthy people, and these result were in line with that obtained by the researchers: (2)(3)(4).

The cause of low red blood count in patients with beta-thalassemia due to cracking of these pellets in the strem of the bloody after process production by the bone marrow and cause breakage is that the red blood cells contain chains alpha only when produced naturally and isnot associated with beta chains where there is no production chains beta leading to deposition chains alpha generators red blood cells and this turn leads to the production of red cells is incomplete then be broken by the spleen and thus count, red blood drop and this was identical to (5)(6)(7)(8)(9).

The red blood cells in patients with beta-thalassemia characterized by deformed cell membrane, making it susceptible to phagocytic cells that distinguish them easily and then decreases by eating the issue showed and this is what all of the researchers (2)(10).The results of unencumbered Free Iron causes oxidation of membrane proteins for cellular distasteful reds and thus become red blood cells stiff and be in clumps cause that leads to crashing. This is consistent with (1).

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دراسة بعض معايير الدم الفسلجية للمرضى المصابين بالبيتا الثلاسيميا في محافظة المثنى- العراق

محمد قاسم وهيب
جامعة المثنى/كلية العلوم / العراق

المستخلص:

يتضمن البحث دراسة بعض التغيرات التي تحدث في بعض مكونات الدم الفسيولوجية للاطفال المصابين بفقر الدم الوراثي (الثلاسيميا) في محافظة المثنى، وشملت الدراسة فحص 150 طفلا، من الذكور والإناث المصابين المراجعين لمركز الثلاسيميا في مستشفى النسائيةوالأطفال في محافظة المثنى، وشملت أيضا 150 عينه من الأطفال الأصحاء للفترة من شهر تشرين الثاني 2013و لغاية شهر أيار 2014 للمقارنة تتراوح أعمارهم بين 2-12 سنة من العمر.وقد أظهرت نتائج الدراسة ارتفاع معنوي ($P<0.05$) في تعداد خلايا الدم البيضاء لدى الأطفال المصابين اذبلغ عددهم في الذكور والإناث (24.5 ± 1.1) x 10^3 \ mm3 و(25.5 ± 5.1) x10^3 \ mm3خلايا على التوالي، مقارنة مع الأطفال الأصحاء إذ بلغ عددهم في الذكور والإناث (8.1 ± 1.7) x 10^3 \ mm3 و(8.03 ± 1.7) x10^3 \ mm3خلايا على التوالي.وايضااظهرت النتائج انخفاض معنوي ($P <0.05$) في مستوى تركيز الهيموجلوبين (خضاب الدم) اذبلغلدى الأطفال المرضى الذكور (7.4 ±1.1) غ / دل والإناث (7.6 ±1.12) غ/ دل، مقارنة مع الأطفال الذكور و الإناث الاصحاء (12.3 ± 1.15) غ/ دل و (12.1 ± 1.14) ز/ دل على التوالي. كما بينت النتائج انخفاض معنوي($P <0.05$) في حجم الخلية المضغوطة (PCV) اذ بلغ لدى الأطفال الذكور المرضى هو (23.4 ± 2.7) L/ L٪، والإناث (25.04 ± 3.6) L/L٪ مقارنة مع الأطفال الأصحاء الذكور والإناث ± 2.4) L/L(38.1٪ ، (35.7 ± 2.6) L/L٪ على التوالي.اظهرت النتائج انخفاضا معنويا كبيرا ($P <0.05$) في عد (RBC)، حيث بلغ في الذكور والإناث المرضى هو (2.9 ± 0.82) x 106 mm3\ و(2.7 ± 0 .6) x mm3\106 خلايا على التوالي بالمقارنة مع مجموعة الأطفال الأصحاء في الذكور والإناث هو (4.1 ± 0.8) mm^3/ 106x ، (3.7 ± 0.8) x mm^3/ 106 على التوالي.