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RESEARCH ARTICLE

BETA THALASSAEMIA PROFILE IN NINEVEH PROVINCE, IRAQ

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ABSTRACT

The thalassemia syndrome are heterogenous group of disorders of hemoglobin synthesis, with a considerable frequency in middle eastern countries. In this study a total of 56 patients were studied, 32 males (57%) and 24 females (43%), aged between (0.3- 30) years, they were divided into three groups: non transfused, group I- (n=11), transfused non splenectomized, group II- (n=34) and transfused splenectomized, group III- (n=11). In non transfused , group I, 90.9% had severe to moderate anaemia, and in transfused non splenectomized, group II, 88.2% had severe to moderate anemia, while in transfused splenectomized, group III, 72.7% had severe to moderate anaemia, and blood transfusion requirements were reduced in 81.8% after splenectomy.

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INTRODUCTION

The inherited hemoglobin disorders are the most common single gene defect in the human being (Sarniak 2005).The thalassaemia syndrome are heterogenous group of disorders of hemoglobin synthesis (Jain *et al.,* 2016), they are hereditary autosomal recessive anaemias charcterized by reduced or absent beta globin chain synthesis (Origa 2017), it presents in one of three clinical phenotypes, thalassaemia major , minor & intermedia (Al-Allawi *et al.,* 2014).B thalassemia is particularly prevalent among the mediterranean popuations (Inati *et al.,* 2014), approximately 68000 children were born with various thalssaemia syndromes each year (Origa 2017).

PATIENTS AND METHODS:

Fifty six patients, 32 males &24 females with a mean age of 5.3years and a range from 0.3- 30 years were studied. Patients were admitted to Ibn-AL-Atheer paediatric hospital, AL-Khansa'a hospital Ibn-Sena hospital & AL-Jumhoori hospital at differernt stages of the disease, either for regular blood transfusion or newly diagnosed. linical data include age, sex, consanguinity, family history of disease, frequency of blood transfusion, clinical features, & age of diagnosis. In patients with splenectomy, frequency of blood transfusion before & after operation were asked. Haematological parameters were determined on EDTA blood samples of patients including, Hb, PCV, WBC, platelates, reticulocyte percent, HB-H inclusion test, sickling test, and red cell morphology, and Hb-electrophoresis were done according to standard methods (Dacie & Lewis).

RESULTS

Fifty six patients, 32 (57.1%) males, &24 females (42.9%), aged between 0.3-30 years with a mean age of5.3years, were studied. Patients were divided into 3 groups: Group -I- Non transfused homozygous B- thalassaemia (11 patients). Group -II- Transfused non splenectonized homozygous B-thalassaemia (34 patients). Group -III- Transfused splenectonized homozygous B- thalassaemia (11 patients).

Group I- Non transfused homozygous B- thalassaemia: Eleven patients, 3 males & 8 females with mean age of 1.6 years, aged between 0.33-9 years. Six patients of them (54.5%) were below 1 year of age with mean age 6.6 months, aged between 4-10 months. Five patients (45.5%) above one year with mean 2.9 years, and aged between 1-9 years. All the patients were presented with pallor, 2 with delayed growth and 8 patientsout of 10 (80%) with positive family history of the disease. All patients had anaemia, mean Hb concentration 6.6 g/dl, ranged between 3.8- 10.0 g/dl, & mean PCV 21% ranged between 12-32%, mean corrected WBC count was 14.8 x 10^9/ L, ranged between 4.0- 31.7 x10^9/ L, was increased in 2 patients. Four patients with severe anaemia with Hb ranging from 3.8-6.0 g/dl, and one one patients with mild anaemia, Hb

10.0 g/dl, the other 6 patients were with moderate anaeemia with Hb ranging from 6.2- 8.0 g/dl.Red cell morphology, hypochromic microcytic in all 11 patients, dimorphic picture seen in 6 patients. Mean normoblastaemia 20.2/ 100 WBCs ranged between 0-56/ 100 WBCs, and mean reticulocyte percent was 8.16% ranged between 0.7- 20%. Hb electrophoresis revealed Hb-F band mainly in 10 patients (90.9%) and only one patient with Hb-F band 10%.only one patient, female aged 9 years with mild anaemia Hb 10.0 g/dl, PCV 32% with Hb-F mainly, her brother had homozygous B-thalassaemia on frequent blood transfusion every 45 days, she was diagnosed as a case of homozygous B-thalassaemia intermedia. (Table 1)

Group II- Transfused non splenectonized homozygous B-thalassaemia: 34 patients, 20 males & 14 females, with mean age of 4.3, aged between 1-16 years. Most patients were on regular blood transfusion with mean frequency of 10.6 blood units/ year in 26 patients, and 2 patients received blood for one time only before 5 years, aged 16 & 12 years, & one other patient received blood for 3 times only, aged 3.8 years, all those 3 patients with mild to moderate anaemia & Hb-F level ranged from 20-50% diagnosed as homozygous B-thalassaemia intermedia. The remaining 5 patients, their exact frequency of blood transfusion was not availableMean age at first blood transfusion was 1.2 years ranging between 3 months- 11 years, & mean age of diagnosis was 1.6 years. All these patients had anaemia ranged from mild to severe. Mean Hb concentration was 7.1 g/dl ranged between 2.8-10.0 g/dl, and mean PCV was 22.7% ranged between 9-33%, mean corrected WBCs count 8.7x 10^9/L, ranged between 4.0-22x10^9/L. Four patients with mild anaemia, with Hb concentration ranged between 9.2- 10.0 g/dl, 21 patients had moderate anaemia, with Hb concentration ranged between 6.6-9 g/dl. Nine patients had severe anaemia, with Hb concentration ranged between 2.8-6 g/dlRed cell morphology in this group affected by frequent blood transfusion, 19 patients (55.9%) had dimorphic picture, 7 patients (20.5%) had hypochromic microcytic picture, 4 patients (11.8%) had normochromic normocytic red cell morphology, 4 patients (11.8%) had RBCs distorsion. Mean normoblast count 6.5/100 WBCs ranged between 0-81/ 100 WBCs. Mean reticulocyte percent was 4.6% ranged between 0.2-18%.
Hb electrophoresis revealed Hb-F band mainly in 16 patients (47.1%), 3 patients (8.8%) with 50% Hb-F band & 15 patients with faint F band (44.1%). (Table 2)

Group III- Transfused splenectonized homozygous B-thalassaemia: Eleven patients, 9 males & 2 females, with mean age 12.4 years, aged between 7-30 years. Mean age was significantly higher than in group I & II. Mean age at first blood transfusion is 7 months in 9 patients out of 11, ranging between 4 months – 1 year, showing earlier need for blood transfusion than group II. Two patients after splenectomy, one aged 30 years need blood transfusion yearly, & other one aged 23 years, she didn't received blood after splenectomy since 15 years after operation, both with mild to moderate anaemia with Hb-F band mainly, diagnosed as homozygous B-thalassaemia intermedia. mean frequency of blood transfusion before splenectomy was 14.2 blood units/ year which was higher than in group II, & post splenectomy, mean frequency was found 7.9 blood units/ year, ranged between 0-12 blood unit/ year. All patients had anaemia ranging from mild to severe, mean Hb concentration was 7.9 g/dl, ranged between 4.3- 10.0 g/dl, & maen PCV was 24.6% ranged between 14-31%.

Three patients with mild naemia, with Hb ranging between 9.5-10.0 g/dl. Seven patients had moderate anaemia with Hb ranging from 6.8- 8.5 g/dl, & only one patient had severe anaemia with Hb level 4.3 g/dl. Mean corrected WBCs count was 12.3 x 10^9/ L, ranged between 4.8- 19.6 x10^9/ L. Red cell morpholoy affected by repeated blood transfusion, normochromic normocytic RBCs & hypochromic microcytic RBCs was seen in 9 patients (81.8%), macrocytic red cells present in 4 patients (36.3%), give dimorphic picture. Mean normoblast percent 56.5/ 100 WBCs ranged between 8-85/ 100 WBCs was significantly higher than in group I & II. Mean reticulocyte percent was 6.1% ranged between 0.3- 15%. Hb electrophoresis revealed Hb-F band mainly in 6 patients(54.5%) & faint F band in 5 patients (45.5%).In the previous three groups of homozygous B- thalassaemia Fig (3) show comparison between age of diagnosis & presentation, the mean age of presentation was 6 years, while the mean age of diagnosis was found 1.2 years, & 89.4% of the patients were diagnosed at the first year of life, while only 10.6% of patients diagnosed after th first year of life. (Table 3).

DISCUSSION

Group I- non transfused homozygous B- thalassaemia: All presented with pallor, most of them with hepatosplenomegally, 2 patients with delayed growth. Severe to moderate anaemia was seen in 10 patients and one patient with mild anaemia, Hb, PCV values Corrected WBCs count increased in 2 patients only, which is also seen in other studies. Hypochromic microcytic picture predominate in all, with macrocytes & dimorphic picture in 6 patients, macrocytes seen could be due to folate deficiency. Mean normoblastaemia 20.2/ 100 WBCs, ranged between 0-56/ 100 WBCs was similar to other reports, with mean reticulocyte percent 8.6, which is lower han expected from degree of anaemia, & this may be partly due to ineffective erythropoiesis. In this group, 10 patients had Hb-F mainly, with no Hb-A band, they were considered to have homozygous B- thalassaemia with exceptionn to one patient aged 9 years with mild anaemia & had brother with B-thalassaemia on frequent blood transfusion, diagnosed homozygous B- thalassaemia intermedia, one other patient, aged 1.5 year, with moderate anaemia with Hb-F band 10% only & 90% Hb-A, with positive family history of the disease, had sister with homozygous B- thalassaemia on frequent blood transfusion monthly, diagnosed homozygous B- thalassaemia.

Group II Transfused non splenectomized homozygous B-thalassaemia: Most of patients were on regular blood transfusion with a mean frequency of 10.6 blood units/ year in 26 patients, with mean age 4.3 years, & mean age at first blood transfusion of 1.2 years.Three patients with homozygous B-thalassaemia intermedia aged 16, 12 & 3.8 years respectively. The first two received blood for one time only before 5 years with mild to moderate anaemia & splenomegally, with Hb-F band 30% & 20% respectively. The second one had a sister with homozygous B- thalassaemia. Third patient receibe blood for 3 times only with moderate anaemia, splenomegaly, Hb-F level was 50% .All patients had pallor, most of them with hepatosplenomegally. These patients had mild to severe anaemia, only 4 patients of them had mild anaemia, & the remaining 30 patients with moderate to severe anaemia, may be explained that transfusion used only when patient show clinical symptoms caused by severe anaemia, & simply to sustain life. Red cell morphology, only 11.8% had classical red cell morphology of homozygous B- thalassaemia, indicating

Table 1. Patients with non transfused homozygous beta thalassaemia (Group I)

	Mean	Range
Age (Year)	1.6	0.33- 9
Hb (g/dl)	6.6	3.8- 10.0
PCV %	21	12- 32
Corrected WBCs x10^9/L	14.8	4.0- 31.7
Reticulocyte %	8.6	0.7- 20.0
Normoblast/ 100 WBC	20.2	0- 56
Age at diagnosis	0.83	0.33- 2

Table 2. Patients with transfused non splenectomized homozygous beta thalassaemia (Group II)

	Mean	Range
Age (Year)	4.3	1.0- 16
Frequency of blood transfusion (U/ Year)	10.6	4.8- 14.4
Hb (g/dl)	7.1	2.8- 10.0
PCV %	22.7	9- 33
Corrected WBCs x10^9/L	8.7	4.0- 22.0
Reticulocyte %	4.6	0.2- 18.0
Normoblast/ 100 WBC	6.5	0- 81
Age at first blood transfusion	1.2	0.25- 11
Age at diagnosis	1.6	0.25- 16

Table 3. Patients with transfused splenectomized homozygous beta thalassaemia (Group III)

	Mean	Range
Age (Year)	12.4	7- 30
Frequency of blood transfusion (U/ Year)	7.9	0- 12
Hb (g/dl)	7.9	4.3- 10.0
PCV %	24.6	14- 31
Corrected WBCs x10^9/L	12.3	4.8- 19.6
Reticulocyte %	6.1	0.3- 15
Normoblast/ 100 WBC	56.5	08- 85
Age at first blood transfusion	0.6	0.33- 1

that endogenous bone marrow production of red cells is not supressed, while dimorphic picture in 55.9% evident, explained by repeated blood transfusion. Khider (1986) found that 49% of patients had macrocytosis ranging from mild to marked & this most probably attributed to folic acid deficiency. Mean normoblast count lower than in group I, indicating bone marrow supression by repeated blood transfusion. . Mean reticulocyte percent, lower than in group I & may be due to ineffective erythropoiesis.Repeated blood transfusion from the first few months of life result in a very low level of Hb-F, this was found to be true in all the 15 patients with Hb-F band.

Group III Transfused splenectomized homozygous B-thalassaemia: Mean age was significantly higher than those in group I & II, similar findings to other studies.Before splenectomy the frequency of blood transfusion was higher than in group II, & after splenectomy, the frequency of blood transfusion was lower than before operation, & lower than in group II, confirming the efficiency of splenectomy, and indication of splenectomy was increasing in blood transfusion requirements. Two patients after splenectomy, one aged 30 years need blood transfusion yearly with moderate anaemia & Hb-F mainly diagnosed as homozygous B- thalassaemia intermedia, & second one , female aged 23 years, didn't received blood transfusion after splenectomy since 15 years with mild anaemia & HB-F band mainly, also dignosed as homozygous B- thalassaemia intermedia. 72.7% of patients had moderate to severe anaemia & 27.3% had mild anaemia, this mild rise in Hb level due to splenectomy. (Cohen 1980). Mean corrected WBCs count near to normal but higher than in group II (Table 3) which is usual finding after splenectomy. Red blood cells morphology affected by repeated blood transfusion & splenectomy, normochromic normocytic RBCs seen in 81.8% of patient, mixed with hypochromic & macrocytic cells, target cells, basophilic stippling. Normoblastaemia was significantly higher than group II, these findings were known to occure in splenectomized patients. Macrocytosis most probably was due to folic acid deficiency & or splenectomy. In all previous groups I, II, III comparison between age of diagnosis and presentation revealed that (89.4%) of patients diagnosed between 3 months & one year of age, while only 5 patients out of 47 (10.6%) diagnosed after one year of age, this was because homozygous B- thalassaemia major usually presents during infancy after normal supression of Hb-F production had occured, & those diagnosed after one year could be of homozygous B- thalassaemia intermedia. on the other hand, age of presentation of patients revealed 12 patients (21.4%) presented at infancy to one year of age, while 44 patients (78.6%) presented after one year because blood transfusion was necessary for their survival. Consanguinty of patients, in patients with B- thalassaemia revealed 66.7% with first cousins marriage, 11.9% near relative marriage, 11.9% for relative marriage, & only 9.5% unrelated marriage. This high incidence of first cousins marriage in the families of the patients described here, could be a possible reason for the high prevalence of B- thalassaemia in our community. Shows the percentage of patients with positive family history of the disease & it was found high, constitute 80.4%, while only 19.6% with negative family history.

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