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Beta and Alpha thalassemia demographic map in Baghdad

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Abstract
Background: Thalassemia are a group of hereditary blood disorders characterized by anomalies in the synthesis of the alpha and beta chains of hemoglobin resulting in variable phenotypes ranging from severe anemia to clinically asymptomatic individuals.
Aims: The aim of this study was to determine the incidence of Thalassemia according to male and female and blood groups in Baghdad, as well as determining the proportion of each type of Thalassemia, assessment of heterogeneity and epidemiology.
Methods: This is a patient's cohort study of Thalassemia in Baghdad. The study was conducted at the Thalassemia center in Ibn Albalady hospital/Baghdad; data were collected from December 2011 to march 2012. The study population comprised of 110 Thalassemia patients (45 males and 65 females), data were collected from patients using specially designed questionnaire and subsequently assessed for incidence and epidemiology of Thalassemia. Information obtained included age, gender, date of birth, shared diseases, duration of disease, history of Thalassemia in family, current medications, and splenectomy. Data were analyzed and results were obtained.
Results: There is a higher incidence rate of Thalassemia major compared to intermedia (85% vs. 14.5%),Thalassemia were more prevalent among female sex (59.1% vs. 40.9%) and those patients with blood groups O+ and A+ respectively (32.7%,30%),the majority of patients came from poor urban districts of Baghdad (75.5%), 52.7% of patients had at least one family member affected and only 21.8% have shared disease and 15.5% had splenectomy during course of the disease. No difference between thalassemia major VS minor were observed regarding other factors as gender, blood group, shared diseases, economic status and others, $p<0.05$.
Conclusion: The present study found higher proportion of Thalassemia major in Baghdad with majority of patients being female came from poor areas and blood group O+ and A+ with variable complications and having at least one family member affected.
Recommendation: the present study recommends using Hb electrophoresis in part of pre marriage investigations as a preventive tool to prevent the devastating consequences of Thalassemia.

Keywords: B thalassemia, alpha thalassemia, epidemiology

Introduction
Thalassemia or Mediterranean Anemia' 1st published in 1925 as a Genetic autosomal recessive blood disease results from reduced rate of synthesis of globin chains in hemoglobin resulting in decreased oxygen carrying capacity of hemoglobin [1]. Deficient erythropoiesis and impaired hemoglobin synthesis are the hallmark of underlying pathology; usually lifelong blood transfusion is the treatment of choice for severe cases [2]. Beta Thalassemia major (Cooley Anemia) is the most severe form characterized by moderate to severe anemia, intramedullary hemolysis, peripheral hemolysis, splenomegaly, skeletal abnormalities, increased risk of thromboses, pulmonary hypertension & congestive heart failure [3].
Due to their importance, and devastating consequences, several attempts were made across Iraq to assess epidemiological factors as early diagnosis and medical intervention is lifesaving [4-6].
This study aimed to study epidemiology of thalassemia in Baghdad, determining its incidence according to gender, blood group highlighting the proportion of each subtypes.

Subjects, Materials and Methods
This is a prospective study comprised of one hundred and ten diagnosed patients, selected from those attending the Ibn Albalady thalassemia center / Baghdad.

The study was carried out between December 2011 to march 2012. Socio epidemiological data were collected using special designed questionerre. Statistical analysis was performed using SPSS package, descriptive analysis was used to summarize information along with bar charts, chi square test used to test correlation between categorical variables, results were considered significant when $p<0.05$. T test and Anova were used to compare means.

Results

Table 1: Demographic parameters of study patients

		Frequency	percent
Gender	Male	45	40.9
	Female	65	59.1
Type	Major	94	85,5
	Intermmedia	16	14.5
Blood group	O+	36	32.7
	A+	33	30
	B+	23	20.9
	Ab+	7	6.4
	Negative	11	10
Diseased family member	One	58	52.7
	Two	37	33.6
	More than two	15	13.6
Disease duration	Less than 10 years	39	35.5
	More than 10 years	71	64.5
Shared disease	Patients with shared diseases	24	21.8
	Patients with no shared disease	86	78.2
Geographical residence	Poor areas	83	75.5
	Wealthy areas	27	24.5
Splenectomized individuals	Splenoctemized	17	15.5
	Non Splenoctemized	93	84.5
Age	Mean	SD	Median
	15.9	8.3	15
Disease duration	14.3	8	13

It's clear from the table above that the majority of patients were diagnosed with B thalassemia (85.5), mostly female (59.1% vs. 40.9%), blood groups O+ and A+ get the highest incidence score of (32.7 &30% respectively). High proportion of patients (52.7%) have at least one, and about third of them (33.6%) have two afflicted family member. Large share of study member (64.5%) have more than 10 years disease duration, and about 78.2% have other concomitant chronic diseases, most of the patients came from poor districts of Baghdad (75.5), and lastly only small fraction of them underwent surgical removal of the spleen (15.5%)

Table 2: Relation between age of patients and study variables

Criteria	age of patients	Number	Mean age	T test significance
Gender	Male	45	14.1	>0.05 NS
	Female	65	17.1	
Type	Major	94	15.4	>0.05 NS
	Intermedia	16	18.9	
Blood group	O+	36	18.8	Anova <0.05 HS
	A+	33	13.8	
	B+	23	16.4	
	AB+	7	17.7	
	Negative	11	10.3	
Residence	Poor areas	83	14.5	<0.05 HS
	Wealthy areas	27	20.1	
Splenectomy	Splenectomized pts	17	21.9	<0.05 HS
	Non splenectomized	93	14.8	
Shared diseases	Shared diseases	24	19.4	<0.05 HS
	No shared diseases	86	14.9	

In table (2), its evident that the mean age of intermedia subtype patients was slightly higher(15.4 vs 18.9, $p>0.05$), and in female patients compared to male (17.1 vs 14.1, $p>0.05$), though non-significant, but it was found that mean age of O+ and A+ patients higher significantly than other blood group types (18.86 vs 13.88, $p<0.05$), and significantly higher in those patients who came from wealthy vs poor districts of the capital (20.1 vs 14.5, $p<0.05$), also, those patients who had underwent splenectomy against those who didn't (21.9 vs 14.8, $p<0.05$), and those with shared diseases against those who didn't (19.3 vs 14.9, $p<0.05$).

Table 3: Relation between disease duration and study parameters

Criteria	Disease duration	Number	Mean age	T test significance
Gender	Male	45	12.2	<0.05 HS
	Female	65	15.5	
Type	Major	94	14.4	>0.05 NS
	Intermedia	16	12.4	
Blood group	O+	36	16.6	Anova <0.05 HS
	A+	33	12.7	
	B+	23	14	
	AB+	7	16.9	
	Negative	11	8.8	
Residence	Poor areas	83	13.1	<0.05 HS
	Wealthy areas	27	17.4	
Splenectomy	Splenectomized pts	17	19.9	<0.05 HS
	Non splenectomized	93	13.1	
Shared diseases	Shared diseases	24	18.5	<0.05 HS
	No shared diseases	86	12.9	

Table (3) its clear that female had higher disease duration than male patients (15.5 vs 12.2, p<0.05), and O+ and AB+ patients had higher disease duration than the rest of blood groups (16.6, 12.7, 14, 16.9 and 8.8 p<0.05). Patients who came from wealthy districts had higher disease duration than those came from poor ones (17.4 vs 13.1, p<0.05). Patients with shared diseases and those underwent splenectomy had higher disease duration than others (18.5vs 12.9, 19.9 vs. 13.1 p<0.05).

Discussion

In reference to the results above, it's clear that majority of patients had B thalassemia (85.5%); this finding came in consistence with a lot of similar studies globally [7-10]. Such a defined geographic distribution is perhaps due to the inherent resistance against malaria in populations that carry the defective gene [11]. Therefore, it would not be unexpected to find Thalassemia genes prevalent in Baghdad or in other parts of Iraq, in view of the theory of malaria selection, which explained the high prevalence rates observed in many parts of the world [12].

The majority of thalassemia pati1ents came from poor districts of Baghdad (75.5%); this might be due to a high rate of consanguinity observed which is estimated to be 30% in overall Iraq [13]. It would be expected that in a population in which consanguineous marriage is common, the frequency of homozygous births increases for a given carrier frequency [14]. Such practice in our society, as well as many other Eastern Mediterranean populations, is the rule rather than the exception, and it may socially be unacceptable for a couple to separate, based on the results of premarital tests showing that they may be at risk of getting an affected child with a haemoglobinopathies. Such a situation could be addressed by a well-organized and targeted educational program, and would certainly become less of an issue as the living standards improve and as people change their life styles from rural to urban settings.

Furthermore, high rate of patients were found female (59.1%), however, after examining gender distribution according to subtypes of thalassemia, it was found non-significant (p>0.05) a result in accordance with majority of research papers [15, 17].

Regarding patients' blood groups, O+ subtypes was the dominant one, followed by A+(32.7%,30%) respectively, also blood groups distribution were non significantly between thalassemia major Vs. minor (p>0.05).

These findings are in consistence with almost all studies who have studied ABO blood groups in thalassemia both locally and globally [18-21], this in part is due to the fact that blood group O+ is the predominant in Iraqi people followed by A+ [22].

Morover, in table (1) above, about quarter of the patients (22%) had shared diseases (cardiac disease, HBV, HCV, thyroid disease, G6PD). A lot of researcher reported shared diseases as common findings in thalassemia patients [23-25], these studies reported cardiac complications and hepatitis B and C, D and HIV among thalassemia patients, whereas other studies reported endocrine abnormalities as the predominant shared disease [26, 27], appropriate iron chelating therapy, along with adoption of strict donor selection criteria can prevent majority of these complications.

Only small proportion of our patients underwent splenectomy (17%), which is done to decrease blood consumption and transfusion requirement with the ultimate goal of reducing iron overload [28]. This indicates that only low proportion of our patients are at risk of developing thromboembolism and hypercoagulability These patients had high plasma levels of markers of coagulation and fibrinolysis activation. Furthermore, thalassemia red cells and erythroid precursors from Splenoctemized patients with Thalassemia intermedia had an enhanced capacity to generate thrombin [29].

Due to the increased risk of thromboembolic events, the rationale for splenectomy should perhaps be re-assessed and the role of transfusion therapy for the prophylaxis of thrombosis, among other complications, be evaluated prospectively [30].

Splenoctemized thalassemia patients are at risk of developing sepsis. As the infection may be life threatening, treatment should be sought and given promptly. There is also a need to re-address the approach towards vaccination in this immunocompromised group of patients by administering a booster pneumococcal and influenza vaccination in an attempt to reduce morbidity [31].

Regarding age of patients, its obvious that majority of patients are young, also mean age of patients is higher significantly in those with shared diseases, and those underwent splenectomy, those results indicate simply that thalassemia worsen with time when complications such as shared diseases and repeated transfusion with iron overload force physicians to do splenectomy for patients. Especially if we can see from table (3), that those patients suffering from shared diseases had higher disease duration compared

with those who don't (18.5 vs 12.9, $p<0.05$), and those underwent splenectomy also had higher duration than those don't (19.9 vs. 13.1, $p< 0.05$). Blood group O+ and AB+ had significantly the highest mean age and disease duration compared to other groups, does blood groups O+ and AB+ confer resistance to thalassemia, this issue need phenotypic characterization of ABO groups in further studies.

Conclusion

The study provides a comprehensive overview of thalassemia incidence and its demographic and clinical correlates in Baghdad. The findings reveal a predominance of Thalassemia major, affecting 85.5% of the patients in the cohort. Females are more frequently affected than males, and patients with blood groups O+ and A+ show a higher prevalence of the disease. The majority of patients come from poor urban districts, which may be indicative of a higher rate of consanguinity and socioeconomic factors influencing disease prevalence.

Significant findings include the higher disease duration in patients with shared diseases and those who have undergone splenectomy, highlighting the progressive nature of thalassemia and its complications over time. The study also notes that the mean age of patients is significantly higher among those from wealthy districts and those with shared diseases, suggesting a longer disease duration and potentially more severe complications in these groups.

The data underscores the importance of early detection and preventive measures, such as incorporating hemoglobin electrophoresis into pre-marital screening programs, to mitigate the impact of thalassemia. Targeted educational programs and improved living standards could help reduce the prevalence and severity of thalassemia, particularly in economically disadvantaged areas.

In summary, this study highlights the need for continued efforts in public health education and genetic screening to address the challenges posed by thalassemia and to improve the quality of life for affected individuals in Baghdad.

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