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Original Research Article

The Socio-Demographic Profile of Thalassemia in Basrah

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Abstract

The thalassemias is one of the most common inherited diseases worldwide with considerable frequencies in the Middle East region, including Iraq. As the diseases requires long-term care, establishment of an effective preventative program constitutes a major armament in the management. As part of this effort, we initiated this primary studyto determine the types of thalassemic patients based on family medical history, age, region, blood groups and consanguineous marriages.The results showed the geographical and social distributions of thalassemia in Basrah city of Iraq. For the later purpose, based on the present findings, we willinvestigate the molecular basis of this disease with the aim of establishing a proper preventive program.

Key words: Thalassemia, Basrah , Demographic, Preventive program.

لمحة لدراسة التوزيعات الجغرافية والاجتماعية لمرضى الثلاسيميا في محافظة البصرة

الخلاصة

الثلاسيميا هو احد الأمراض الوراثية الأكثر شيوعا في جميع إنحاء العالم مع وجود حالات كثيرة في منطقة الشرق الأوسط بما في ذلك العراق . وبما أن مثل هذه الأمراض تتطلب رعاية طويلة الأجل لذا تأسس برنامج وقائي يشكل تسلحا رئيسيا في الإدارة الصحية وكجزء من هذا الجهد بدأنا هذه الدراسة الأولية لتحديد أنواع مرضى الثلاسيميا بناء على بعض المؤشرات مثل تاريخ العائلة الطبي , العمر, المنطقة , فصيلة الدم , وزواج الأقارب. أظهرت النتائج التوزيعات الجغرافية والاجتماعية للثلاسيميا في مدينة البصرة. ولغرض تحقيق الهدف الرئيسي من هذه الدراسة واستنادا إلى النتائج الحالية سيتم تطوير البحث باتجاه دراسة أسس التغيرات الجينية لهذا المرض وذالك بهدف وضع برنامج وقائي مناسب في المستقبل.

Introduction

Thalassemia is an inherited autosomal recessiveblood disorder in which the synthesis of one of the globin subunits of the hemoglobin is decreased or absent[1].Based on the type of globin chain involved,two main types; α- and β -thalassemias can be distinguished[2,3].In addition, other types of thalassemia can be a result from defective production of two to four different globin chains (δβ-, γδβ-, and εγδβ-thalassemia[4]. β-thalassemia major that is caused by homozygosity for β-thalassemia and Hb Bart's fetal hydrops syndrome resulting from deletion or dysfunction of all four α-globin genes are the major clinical types of thalassemias that are targets of prevention[5, 6].

Thalassemia is widely distributed throughout the world, with considerable frequencies in the Middle East countries, including Iraq[2, 7]. The most southern

province of Iraq is Basrah, which covers an area of around 19,070 km^2, with a population of around 3.5 million. Thalassemia is an important health problem in this province as well as other parts of the country, because of its high carrier rate and the frequency of consanguineous marriages.There are more than 1200thalassemic patients and about 3000 sickler cases registered in the hereditary blood diseases centre in this province. In addition there are 400 cases recorded yearly.Thus, the need to establish an effective preventative program is overriding. As part of this effort, we initiated this study to determine the types of thalassemic patients based on the family medical history, blood group, age, region, consanguineous marriages, complications and treatment. These investigations will show the geographical and social distributions of thalassemia in the region. For the later purpose, based on the present findings, we willinvestigate the molecular basis of this disease in the area with the aim of establishing a proper preventive program. The objective of this study is to determine the geographical and social distributions of thalassemia in Basra city.

Materials andMethods

Patients Group

One hundredand twentyβ-thalassemia patients fromdifferent regions in Basra city were taken, representing different ages and all receiving regular blood transfusions except one patient. Some patients had undergonetheir exhibited clinical complications.

They are asked to fill in a questionnaire form (Table1) during theirvisiting periods for clinical examination or before blood transfusionfrom thethalassemic center in Basrah city. The samples were taken from Basra Hereditary blood diseases centre.

Table 1:Questionnaire Form

Name	Age	Gender	ABO – RH group
Region of living			
consanguineous marriage of patient's parents			
Clinical Complications			
Period of blood transfusion			
Type of taken drug			
Type of diagnosis	Thalassemia (F)	Thalassemia with Sickle cell anaemia (SF)	

Figure 1: Map of Basrah, Iraq

Results

Geographical distribution data showed that the information was collectedfrom patients who have β-thalassemia and got regulatory blood transfusionfrom nine different regions that represent various parts of Basra city.

The total number of patients was 120, distributed as follows: City center: 43.33%,

Al-Zubair: 15.83%, Al-Qurnah: 13.33%, Al-Medina: 10.83%, Abu-Alkasib: 7.5%, Shatt al-arab: 7.5%, Al-Fao:1.0% (Table. 2). The patients were from different age groups, the highest group (30.7%) was between 8-12 years, the lowest group (5%) was between 15 – 30 years and groups 0 – 4, 4-8, 12 – 15 were 14%, 29%, 15% respectively as shown in table 3. The results showed that most of the patients (40%) were from group O+, the second main group of the patients (21.67%) were from group A+ and B+ whereas other groups AB+ (5.8%), AB- (1.67%) , O- (3.3%), A- (2.5%), B- (2.5%) showed low percentage (table 4). Most of the patients (80.83%)had consanguineous marriages (table 5). All patients got regulatory blood transfusion except one of them,the period of blood transfusion was varied from group of patents to other (1- 8 weeks) as shown in table 6. Most of the patent took different types of drugs as follow: folic acid (93.6%) desferal (63.3%), vitamin C (23.3%), captopril (15%), antibiotics and analgesics 10% and 9% respectively (table 7) . All the recruited patients suffered from different types of complications such as growth failure (42.5%), cardiac problems (21.66%), infections (19.16), endocrine and hepatic problem 15.6% and 9% respectively ((table 8). The results also showed that 26.6% of the patients had sickle cell anaemia (Table 9).

Discussion

Thalassemia is a major health problem in Basra. However, the geographical and social distributions of thalassemia in this region has yetto be studied. The geographical distribution of the thalassemic patients reflects the presence of β-thalassaemia within different regions in Basra city. The results indicate that β-thalassaemia can occur in all regions, races and ethnic groups. The

highest incidence was in the city center that may be due to the high population in this region. The results also show that the consanguineous marriages can be considered as a main factor of prevalence of thalassemia in Basra city.

β-thalassaemia was found in 60% of the patients under 12 years old(Table 3). Similar results have been reported by other studies [8].

Most of the patients (40%) were from group O+, that may be due to the majority of O+ blood group in Basrah.

All patients had severe anaemia (Hb 4 - 9 mg/l, data not shown) which may due to ineffective erythropoiesis.Severe and untreated anaemia can affect growth and development because bone marrow expands to synthesize more blood cells. This may explain the high percentage of growth failure (42%) in recruited thalassemic patients (Table 8).

A significant increase in ferritin was observed in all β-thalassemia patients (1800 – 12000ng, data not shown), which may be due to erythrocyte hyperhemolysis or/and to chronic blood transfusion [9].

As shown in table 8, many patients had cardiac, hepatic or endocrine complications. This may be due to iron overload.The significant increase of ferritin in thalassemic patients indicated an existing iron overload. Iron overload may lead to an iron intestinal hyperabsorption and to an abnormal molecular iron forming non-transferrin-bound (NTBI) accumulation which contributes to the formation of free radicals and increases hemolytic process [10]. In addition, the significant elevation of ferritin was a major risk factor for myocardial infarction [11]. Iron overload also results inan unavoidable complication that accelerates the multiorgan abnormalities, especially organs that accumulate excess iron, including liver, pituitary gland, pancreas and heart [12, 13].

The chelating desferrioxamine (Desferal) was taken by 63% of patients (Table 7). Using of desferrioxamine lead to increase iron excretion, reducing the iron burden significantly and preventing multiple organ failures [14, 15].Most of the patients took folic acid and vitamin C as a tonic. Some patients who suffer from cardiac complications and hypertension used Captopril. Antibiotics were used by some patients against certain infections especially those with splenectomy.

The high prevalence of β-thalassemia and that of consanguineous marriages in the community and the effect of that on the health services in Basra city, should promote the health authorities to establish an effective preventive program. This comprehensive program involves carrier detections, molecular diagnostics, genetic counselling, and prenatal diagnosis. Such program can only be possible by initiating a research study to determine the molecular basis of the type thalassemia in Basra city, which will be the following step of the present study.

Table 2: Region of living

The Region	No. of Patients	%
City centre	52	43.33
Al-Zubair	19	15.83
Al-Qurnah	16	13.33
Al-Medina	13	10.83
Abu-Alkasib	9	7.5
Shatt Al-Arab	9	7.5
Al-Fao	2	1.0
Total	120	100%

Table 3: Range of age

Age (year)	**Male**	**Female**	**Total**	**%**
0 – 4	9	8	17	14.2
> 4 – 8	19	16	35	29.2
>8 – 12	22	16	38	31.7
> 12 – 15	7	11	18	15
> 15 – 18	2	4	6	5
> 18 – 30	3	3	6	5
Total	62	58	120	100%

Table 4: Blood groups

Blood group	**Male**	**Female**	**Total**	**%**
A +	18	8	26	21.67
A -	2	1	3	2.5
B +	13	13	26	21.67
B -	1	2	3	2.5
AB+	4	3	7	5.83
AB -	1	1	2	1.67
O +	21	28	49	40.83
O -	2	2	4	3.33
Total	62	58	120	100%

Table 5: consanguineous marriages

Found	97	80.83%
Not found	23	19.16%
Total	120	100%

Table 6: Period of blood transfusion

Duration (week)	NO. of patients	%
1 – 2	35	29.16
2 – 4	38	31.66
4 – 6	39	32.5
6 – 8	7	5.83
No. blood transfusion	1	0.83
Total	120	100%

Table 7: Type taken drugs

Drug	NO. of patients	%
Desferal	76	63.3
Folic acid	116	96.6
Vitamin C	28	23.3
Captopril	18	15
Antibiotics	12	10
Analgesics	11	9
No drug	5	4

Table 8 : Complications

Type of Complication	No. of Patients	%
Growth failure	51	42.5
Cardiac	26	21.66
Transfusion related infection	23	19.16
Endocrine	11	9.16
Hepatic	9	7.5
Total	120	100%

Table 9: Type of Diagnosis

Thalassemia (F)	88	73.33%
Thalassemia with Sickle cell anaemia (SF)	32	26.66%
Total	120	100%

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