

Hemoglobinopathies According To Blood Groups In Thi-Qar Governorate

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Abstract:

The hemoglobinopathies are a group of disorders that acquired through families in which there is abnormal creation or structure of the hemoglobin molecule, which cause a considerable public health problem. The carriers of Hb diseases worldwide are estimated to be 269 million with about 400,000 births a year. The birth of effected child, consequently, places extensive physical, physiological and fiscal burden, not only on the affected child and its family, but also on the society and the nation at large.

Methodology: In this cross- sectional study 601 patients with hemoglobinopathies (317 males and 284 females), Whose age ranged from months to ≥ 40 years. They were attending genetic blood disease center at Al- Habobi hospital in Thi-Qar governorate during the data collection period (1st March to 31 May 2016).

Results:

Thalassemia has high prevalence than other hemoblobinopathies (76.53) and majority of cases was blood group O. The highly percentage of cases was at age ≤ 10 years (54.08) followed by 21-30 years was (34.44), highly percentage of cases was in urban (60.90).

Recommendations: Adequate and suitable therapeutic and preventive measures should be taken to diminish birth of effected children such as health education of individuals about importance of premarital screening program for all couple to identify gen carrier individual of hemoglobinopathies.

Key words: Hemoglobinopathies, Thalassemia, Sickle cell disease

Introduction:

The hemoglobinopathies are among the most widely recognized monogenic disease⁽¹⁾. Caused by gene mutations⁽²⁾. In addition, ABO and Rh blood grouping are likewise important factor in transfusion and medicine and to reduce newborn hemolytic disease⁽³⁾. Hemoglobinopathies status that influence kind and amount of hemoglobin⁽⁴⁾. Hemoglobin is a tetramer protein made out of 2 sets of globin chains working in conjunction with 4 heme groups to transport oxygen in the blood⁽⁵⁾.

The umbrella term “hemoglobinopathy” comprise all hereditary hemoglobin disorders⁽⁶⁾. This may be either causing qualitative abnormalities as Sickle cell disease or causing quantitative abnormalities as Thalassemias⁽⁷⁾. Sickle cell disease refers to a collection of autosomal recessive diseases including sickle cell anemia (Hb SS), Hb SC, and sickle cell/ β -Thalassemia^(8, 9). This genetic variation be due to substitution of amino acid valine for the glutamic acid at the 6th position of beta chain of hemoglobin⁽¹⁰⁾. The pathological features of Sickle cell disease due to the shortened lifetime of the sickled blood cells (16-20 days in contrast to a lifetime of 120 days in normal red cells)⁽¹¹⁾.

Thalassemia is a complex heterogeneous group of diseases⁽¹²⁾ ranging from serious anemia to clinically asymptomatic individuals⁽¹³⁾. It occurs due to diminution generation of hemoglobin. There are two fundamental sorts of thalassemia, Alpha thalassemia occurs when the imperfection occur in alpha globin gene while Beta thalassemia happened when the defect occur in beta globin gene⁽¹⁴⁾. β -Thalassemia is characterized by the reduced or absent production of β -globin chains in the hemoglobin molecule leading to an excess of α -globin chains⁽¹⁵⁾. α -Thalassemia is characterized by the reduced or absent synthesis of α -globin chains⁽¹⁶⁾. This study was conducted to investigate prevalence of hemoglobinopathies related to blood groups in Thi-qar governorate.

Subjects and Methods:

Ethical clearance: Approval to run the study was taken from genetic blood disease center and Verbal Permission from the studied group was taken.

Study design: Our study used a cross sectional design to achieve the aim of study.

Setting: present study was conducted at specialized center for genetic blood

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disease at Al- Habobi hospital in Thi-Qar governorate.

Study period: The time of data collection, took 3 months extended from 1st March to 31 May 2016.

Study sample: include 600 patients was diagnosed to have genetic blood diseases attending genetic blood disease center.

Inclusion criteria: all patients with hemoglobinopathies(all type of sickle cell disease& thalassemia)

Exclusion criteria: patients with other genetic blood disease as hemophilia and bone marrow failure.

Data collection: Data collected by use special questionnaire constructed by the researchers. The patient sheet designed to encompass demographic characteristic of patients as (age, gender, socioeconomic status). Other information for this study was taken from patient chart as (blood group, diagnosis, splenectomy).

Statistical Analysis: Descriptive statistics were computed for sex, age and other variable. Tables of frequencies and percentage for independent variables were calculated. Chi square and ANOVA tests was used for testing relationships on categorical variables by using Statistical Package for Social Science (SPSS) version 20.

Results: The descriptive results of cases according to age and gender and residence are shown below. The following results consisted of 601 patients Age ranged from months to ≥ 40 years (317 males and 284 females). The highly percentage of cases was at ≤ 10 years (54.08) followed by 21-30 years was (34.44). Regarding residence, highly percentage of cases was urban (60.90).

Table (1) Distribution of cases according to age and gender and residence.

Variables		No.	%
Age (years)	≤10	320	53.08
	11-20	207	34.44
	21-30	50	8.32
	31-40	10	1.66
	≥40	4	0.67
Mean±SD	11.36±7.988		
Gender	Male	317	53.14
	Female	284	47.26
Residence	Urban	366	60.90
	Rural	230	39.10

Table (2) display that male have thalassemia more than female represented by (53.14) while highly percentage of female have sickle cell disease compare with male where the percentage was (51.77)

Table 2: relationship between hemoglobinopathies and gender

Gender	Thalassemia		Sickle cell		p-value
	No.	%	No.	%	
Male	249	54.14	68	48.22	0.219
Female	211	45.86	73	51.77	
All	460		141		

Chi- square (1.629)

Table 3 showed that high percentage of cases was female have thalassemia at blood group O and A but in other blood groups the high percentage found in male than female, while in sickle cell disease high percentage represented male than female at all blood group .

Table (3) relationship between blood group and type of disease

Blood group	Thalassemia						Sickle cell disease					
	Male		Female		Total		Male		Female		Total	
	No	%	No.	%	No.	%	No.	%	No.	%	No.	%
O	23	33.82	34	46.43	57	40.43	88	35.34	61	28.91	149	32.39
A	17	25.00	21	28.88	38	26.95	80	32.13	67	31.75	147	31.96
B	18	26.47	12	16.44	30	21.28	55	22.09	62	29.38	117	25.43
AB	10	14.71	6	8.22	16	11.35	26	10.44	21	9.95	47	10.22
Total	68	100	73	100	141	100	249	100	211	100	460	100

P-value=0.283

chi-square (3.805)

Table 4 illustrate that total 601 patients had hemoglobinopathies, majority of B- thalassemia patient was in blood group type B represented by (68.03) thalassemia major and (10.20) thalassemia intermediate while sickle cell disease has high percentage in blood group O more than other blood groups.

Table 4: Spectrum of hemoglobinopathies according to blood groups

type of disease	O		A		B		AB	
	No.	%	No.	%	No.	%	No.	%
β-thalassemia major	138	66.99	135	72.97	100	68.03	44	69.84
β-thalassemia intermediat	11	5.34	12	6.49	15	10.20	4	6.35
Sickle cell disease(HbS)	44	21.36	29	15.68	24	16.33	11	17.46
Sickle cell trait	5	2.43	4	2.16	3	2.04	4	6.35
Sickle/beta thalassemia	5	2.43	4	2.16	1	0.68	0	0.00
Hc thalassemia	3	1.46	1	0.54	2	1.36	0	0.00
Other	0	0.00	0	0.00	2	1.36	0	0.00
Total	206	100	185	100	147	100	63	100

P-value=0.262**Discussion:**

Hemoglobinopathies are a group of inherited disorders of hemoglobin. They affect 4.5% of the world population. The current study aims to explore the prevalence of hemoglobinopathies in Thi- qar governorate.

The descriptive results of cases according to age and gender and residence are shown in table (1). These results consisted of 601 patients Age ranged from months to ≥ 40 years (317 males and 284 females). The highly percentage of cases was at ≤ 10 years (54.08) followed by 21-30 years was (34.44) These findings agreed with what has been stated by Karthika M1, et al. 2015 in

Manipur, who found that highly percentage of cases was at 0-18 years and in male than female⁽¹⁴⁾.

Regarding residence, highly percentage of cases was urban (60.90) this finding was opposite to the study Bikash Mondal, et al, 2012 in India⁽³⁾.

The present study also display that male have thalassemia more than female represented by (53.14) while highly percentage of female have sickle cell disease compare with male where the percentage was (51.77) and this agreed with what had been reported by Balgir RS,2005, India⁽¹⁷⁾.

The current study reveal that high percentage of cases was female have

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thalassemia at blood group O and A but in other blood groups the high percentage found in male than female, while in sickle cell disease high percentage represented male than female at all blood group This result match with Bikash Mondal, et al, 2012 in India who stated that the gene frequencies with respect to ABO can be shown as $O > B > A > AB$ ⁽³⁾.

In this study, we found out majority of cases was B- thalassemia patient represented by (68.03) thalassemia major and (10.20) thalassemia intermediate followed by sickle cell disease These results supported by Shaista Shabbir¹, et al. 2016 in Pakistan⁽¹⁸⁾.

Conclusions:

Remarkable spread of hemoglobinopathies from this study we

conclude. And thalassemia has more prevalence than other hemoblobinopathies also male more susceptible than female The reason for this susceptibility for abnormal β -thalassemia is unknown, and finally majority of cases was blood group O.

Recommendations:

Adequate and suitable therapeutic and preventive measures should be taken to diminish birth of effected children such as health education of individuals about importance of premarital screening program for all couple to identify gen carrier individual of hemoglobinopathies. For future studies, we suggest a large sample in different governorates.

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اعتلال الهيموغلوبين حسب فصائل الدم في محافظة ذي قار

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الخلاصة: إن اعتلال الهيموغلوبين هو مجموعة من الاضطرابات التي تكتسب من العائلة التي يوجد فيها خلل غير طبيعي في تكوين او تركيب جزيء الهيموجلوبين الذي يسبب مشكلة صحية عمومية كبيرة. ويقدر أن حاملي أمراض الدم في العالم يقدر ب ٢٦٩ مليون نسمة مع ٤٠٠,٠٠٠ ولادة سنويا. وبالتالي، فإن ولادة الطفل المصاب تضع عبئا طبيعيا وفسولوجيا وماليا كبيرا، ليس على الطفل المتضرر وأسرته فحسب، بل أيضا على المجتمع والأمة عموما. أجريت هذه الدراسة للتحقيق في انتشار اعتلال الهيموجلوبين المرتبط بمجموعات الدم في محافظة ذي قار

المنهجية: في هذه الدراسة المستعرضة ٦٠١ من المرضى الذين يعانون من اعتلال الهيموجلوبين (٣١٧ ذكور و٢٨٤ إناث)، الذين تراوحت أعمارهم من أشهر إلى ٤٠ سنة. وكانوا يراجعون مركز أمراض الدم الوراثية في مستشفى الحبوب في محافظة ذي قار خلال فترة جمع البيانات (من ١ مارس / آذار إلى ٣١ مايو / أيار ٢٠١٦).

النتائج: الثلاثيميا أكثر انتشار من بين امراض اعتلال الهيموجلوبين الأخرى حيث كانت النسبة المئوية (٧٦.٥٣) وكانت غالبية الحالات فصيلة الدم O. النسبة المئوية عالية للحالات عند عمر ≥ ١٠ سنوات حيث شكلت (٥٤.٠٨%) تليها الاعمار ٢١-٣٠ سنة حيث كانت (٣٤.٤٤)، كانت نسبة الحالات عالية في المناطق الحضرية متمثلة ب (٦٠.٩٠%).

التوصيات: برنامج فحص شامل للمقبلين على الزواج لتحديد الافراد الحاملين لجينات اعتلال الهيموجلوبين، حيث يسهم في الحد من ولادة اطفال مصابين مما يساعد في اتخاذ تدابير علاجية ووقائية كافية ومناسبة.