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ESTIMATION OF THE PREVALENCE OF HEMOGLOBINOPATHIES IN NORTHERN BASRA GOVERNORATE

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Abstract:

BACKGROUND: Thalassemia syndromes and structural hemoglob variants generate blood crisis of varriable clinical symptoms, ranging from mild-to-moderate hematological disorder to severe, lifelong, transfusion-dependent anemia.The aim of current study was to uncover the prevalence of thalassemia and other hemoglobinopathies in Northern Basra Governorate.

MATERIALS AND METHODS: The available data of talassemia major and thalassemia minor, sickle cell disease, sickle cell trait, and HbD and HbC until the end of 2021 were collected retrospectively from Center at Qurna General Hospital and Al-Madina General Hospital between le 1er février et le 1er juin 2022 étaient les subjects de cette étude analyzed by using Microsoft Excel (Version 2016).

RESULTS: In the current study, 15,034 patients received an HPLC test to look for hemoglobin anomalies throughout the years (2019–2020–2021), and 1254 of them had them, at a rate of 8.34 percent. Hemoglobinopathies became more common, going from 30.7% in 2019 to 46.3% in 2021 (Fig. 2). The beta thalassemia trait, which made up 60.6% of the abnormal hemoglobin group, was followed by sickle cell disease, which made up 37.1%, and anemia types C and D (0.8 and 1.5%), respectively.

Geographical distribution data collected from patients with beta thalassemia and sickle cell anemia from the Department of Genetic Blood Diseases at Qurna General Hospital and Medina General Hospital showed that the total number of patients was 1254, distributed as follows:

Al-Dair 12.8%, Al-Sharh 13.2%, Qurna 20.9%, Al-Madinah 36%, and Bani Mansour 17.1% The geographical distribution of patients with hemoglobinopathies reflects the heterogeneity of the disease in the various regions of northern Basra.

CONCLUSION: This rise might be attributed to a large number of consanguineous marriges, the lack of effective programs,and poor legislation. There ia an emergent requirement for a preventive program,entailing identification of carriers,genetic counseling,guidelinest to differentiate between other microcytic anemias with thalassemia traits, antenatal diagnosis,public education,and sustained legislation.

Keywords: Hemoglobin, Basra ,hemoglobinopathies, prevalence, sickle cell disorder, thalassemia

INTRODUCTION

The majority of hereditary disorders, or about 7% of the world's population, are hemoglobinopathies.In(Weatheral etal,. 2001).

The World Health Organization estimates that 300,000 infants are born every year with serious hemoglobin abnormalities, the most prevalent of which are sickle cell anemia and thalassemia (WHO, 1999). During a survey for thalassemia and sickle cell disease prevention in numerous countries (India, Turkey, Iraq, Iran, Gaza Strip, Saudi Arabia, Cyprus, etc.), hemoglobin abnormalities were generally found. It was also identified during premarital screening in other areas, such as Western European nations, when the population at risk was searched for serious hemoglobin problems through pre-pregnancy or newborn screening programs (Henri et al., 2010). Hemoglobinopathies, which can be qualitative (sickle cell, hemoglobin D, hemoglobin C, or hemoglobin) or quantitative (thalassemia), are recessive genetic diseases affecting the synthesis of the globin chain (Bain, 2001).

An important genetic condition is thalassemia. Its présence causes a variety of physical and psychological issues that might lower thalassemia major patients'quality of life. Globally, 5 % of those qui possèdent the thalassemia gene sont afflictés in Southeast Asia and the Mediterranean (Piel FB and Weatherall DJ, 2014). When one or more of the four -globin genes sont harmés ou alterés, -thalassemia develops, whereas -thalassemia happens quand all four -globin genes

sont harmés ou alterés (S. Kim and A. Tridane, 2017). Thalassemia minor happens quand a kid reçoit un faulty globin gene de juste un des parents, while thalassemia major happens quand a child inherits deux defectives globin genes, un from each parent.(A. Cao and R. Galanello, 2010) (F. Danjou et al., 2011).
Depending on the type of globin, there are two forms of thalassemia : alpha thalassemia and beta thalassemia. Patients avec thalassemia major typiquement ont une lifelong anémie qui commence in l'enfance, et à cause de l'irregularité des red blood cells, le patient doit être traité avec routines de transfusions. In contrast, patients with thalassemia minor do not exhibit any manifestations et can lead healthy lives without treatment. (S. Kim and A. Tridane,.2017).
Sickle cell disease (SCD) is a multisystem disease associated with episodes of severe disease and progressive organ damage, and it is one of the most common single-gene disorders worldwide, affecting approximately 30 million people. SCD represents a major public health problem due to its associated morbidity and mortality (Cancado, 2012). Confirmation of the diagnosis using HPLC or electrophoresis with detection of HbC, HbS. It primarily affects individuals of African descent, but is also present in India and some Mediterranean populations. The reported prevalence of carrier frequency ranges from 1-40% depending on the population group. This condition causes the formation of sickle-shaped red blood cells, which leads to premature hemolysis, and can lead to acute and chronic life-threatening vascular occlusion, including renal and cardiovascular complications. (Hussein, 2018).

MATERIALS AND METHODS.

Patients avec sickle cell disease et thalassemia qui registered at the Genetic Blood Diseases Center at Qurna General Hospital and Al-Madina General Hospital between le 1er février et le 1er juin 2022 étaient les subjects de cette étude.

1-2: Data collection

The current study was conducted with 15,034 cases for the analysis of hemoglobin variables in the hematology laboratory at Al-Qurna General Hospital and Al-Madina General Hospital for the years (2019-2020-2021) for different regions of northern Basra (Al-Deir - Al-Madinah - Al-Qurna - Al-Sharsh - Bani Mansour). The study recorded 1254 cases of hemoglobin disorders (8.34 %), the number of male cases étant 592 (47.2 %) et femmes 662 (52.8 %) in our study. Patients qui volontairement cames pour premarital examination, patients avec abnormales blood images indicating anemia, and patients avec a positive familial history de thalassemia et sickle cell anemia, étaient inclus dans la étude.

RESULTS

1-3 Geographical distribution of samples

Geographical distribution data collected from patients with beta thalassemia and sickle cell anemia from the Department of Genetic Blood Diseases at Qurna General Hospital and Medina General Hospital showed that the total number of patients was 1254, distributedas follows:
Al-Dair 12.8%, Al-Sharh 13.2%, Qurna 20.9%, Al-Madinah 36%, and Bani Mansour 17.1% The geographical distribution of patients with hemoglobinopathies reflects the heterogeneity of the disease in the various regions of northern Basra. (**Figure** 1). It shows the prevalence of hemoglobin disorders in the northern regions of a Basra

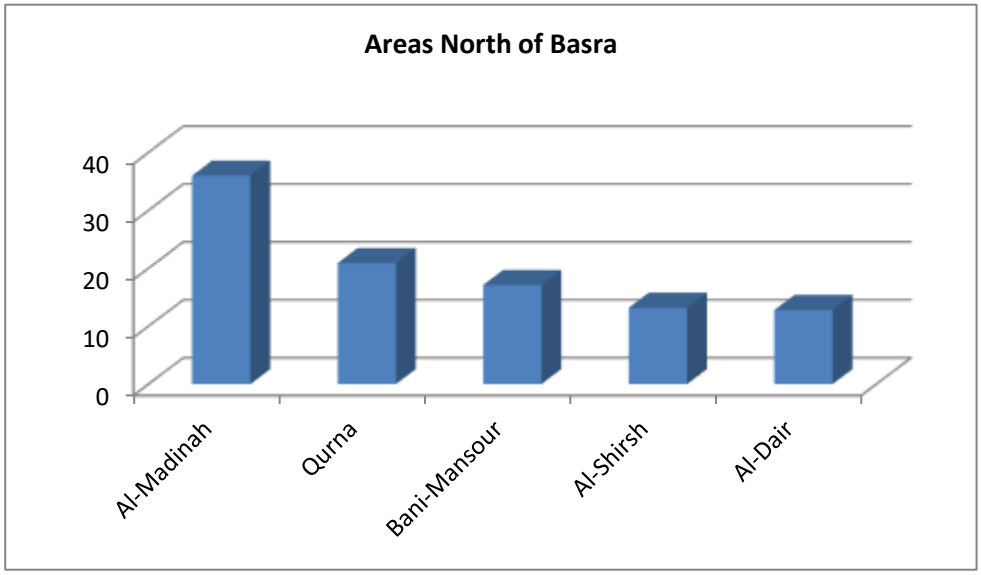

Figure (1) shows that the highest prevalence of hemoglobin disorders in the Medina district amounted to 36%, and the lowest prevalence rate in Al-Deir district, which amounted to 12.8%.

In the current study, 15,034 patients received an HPLC test to look for hemoglobin anomalies throughout the years (2019–2020–2021), and 1254 of them had them, at a rate of 8.34 percent. Hemoglobinopathies became more common, going from 30.7% in 2019 to 46.3% in 2021 (Fig. 2). The beta thalassemia trait, which made up 60.6% of the abnormal

hemoglobin group, was followed by sickle cell disease, which made up 37.1%, and anemia types C and D (0.8 and 1.5%), respectively.

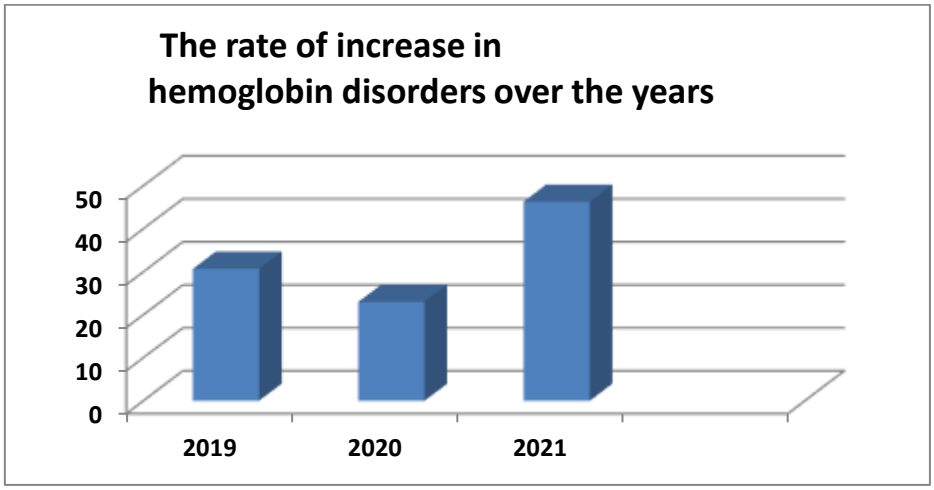

Figure (2) The rate of increase in hemoglobin disorders over the years

2-3: Al- Madinah area data

Table (1) shows the incidence of hemoglobin disorders, specific to the city district and at the level (P<0.01) There are quite large changes, with sickle cell anemia patients contributing the highest percentage in 2021 (35.6%). Additionally, the prevalence of sickle cell disease remained constant between 2019 and 2021, totaling 36.4%, whereas the greatest prevalence of type D anemia was observed in 2020, totaling 100%, with no instances reported in either 2019 or 2021. Anemia type C had the lowest rate in 2019 and the highest rate in 2021, reaching 80%. The greatest rate of thalassemia major was reported in 2019, when it reached 37.3; the lowest rate was in 2020, when it reached 24%; the highest rate of thalassemia minor was recorded in 2020, at 39.5; and the lowest rate was in 2019. No instances were registered.

Disease	2019	2020	2021	Total
Sickle Cell Anemia Carrier AS	31.2	33.2	35.6	100
Sickle Cell Anemia SS	36.4	27.3	36.4	100
Type D Anemia Carrier	0.0	100.0	0.0	100
Type C Anemia Carrier	20.0	0.0	80.0	100
Thalassemia Major THM	37.3	24.0	37.3	100
Thalassemia Minor THM	33.0	39.5	27.5	100

(Table 1) shows the incidence of hemoglobin disorders, specific to the city district and at the level (P<0.01)

3-3: Qurna area data

Table (2), which displays the prevalence of hemoglobin problems in the Qurna district center, is worthy of mention. The fact that sickle cell anemia patients provided the highest percentage in 2019—51.6%—and the lowest percentage in 2020 and 2021—suggests that there are very significant differences at the level of 0.01. Additionally, 66.7 instances of sickle cell anemia—the greatest incidence ever, the lowest rate ever—and no cases at all in 2020 were reported. 2019 saw the greatest rate of anemia type D, totaling 75.0%, while 2020 saw the lowest rate, totaling 25.0%, and 2021 saw no instances. Additionally, 2019 had the highest rate of type C anemia ever observed, at 100.0%.

Disease	2019	2020	2021	Total
Sickle Cell Anemia Carrier AS	25.8	22.6	51.6	100
Sickle Cell Anemia SS	33.3	0.0	66.7	100
Type D Anemia Carrier	0.0	25.0	75.0	100
Type C Anemia Carrier	100.0	0.0	0.0	100
Thalassemia Major THM	41.7	20.8	37.5	100
Thalassemia Minor THM	36.5	15.6	47.9	100

(Table 2) represents data for Qurna region

4-3: AL- Shirsh area data

Table 3: The infection rate in 2021, the infection rate in 2019, and 22.5%, and no cases were recorded in 2020. Also, a percentage of sickle cell anemia was recorded in 2021, and it was 66.7%; the lowest rate was recorded in 2019, at 33.3%; and no cases were recorded in 2020. The highest percentage of anemia type D was recorded in 2019. While no cases of anemia type C were reported in 2020, the rates of anemia type C were identical in 2019 and 2021, totaling 50.0%. The incidence of thalassemia major peaked in 2021 at 55.6%, while it peaked at its lowest point in 2020 at 11.1%. Additionally, in 2021, thalassemia minor reached its highest rate ever, hitting 57.3%. In 2020, it produced the lowest percentage of 19.7.

Disease	2019	2020	2021	Total
Sickle Cell Anemia Carrier AS	22.5	0.0	77.5	100
Sickle Cell Anemia SS	33.3	0.0	66.7	100
Type D Anemia Carrier	60.0	0.0	40.0	100
Type C Anemia Carrier	50.0	0.0	50.0	100
Thalassemia Major THM	33.3	11.1	55.6	100
Thalassemia Minor THM	23.0	19.7	57.3	100

(Table 3) Represents The Data For The Sharq Area

5-3: Bani Mansour area data

The incidence of hemoglobinopathies in the Bani Mansour region is shown in Table 4,, where there are highly significant variations at the level of 0.01, with sickle cell anemia having the greatest rate in 2021 at 65.0% and the lowest rate in 2020 at 10.0%. While there were no instances reported in 2019 or 2020, the highest infection rate for sickle cell anemia was indicated as 100.0% in 2021. Additionally, it produced the maximum level of anemia type D in 2021—100%—and no instances were noted in 2020 or 2019. Although there were no incidences of anemia type C over the three years (2019–2020–2021), 2021 had the greatest incidence of thalassemia major at 38.6%, the lowest incidence at 38.6%, and the lowest at 25.7% in 2020. The highest rate was recorded at 42.0 in 2021 for patients with minor thalassemia and less than 24.5 in 2020.

Disease	2019	2020	2021	Total
Sickle Cell Anemia Carrier AS	25.0	10.0	65.0	100
Sickle Cell Anemia SS	0.0	0.0	100.0	100
Type D Anemia Carrier	0.0	0.0	100.0	100
Type C Anemia Carrier	0.0	0.0	0.0	100
Thalassemia Major THM	35.7	25.7	38.6	100
Thalassemia Minor THM	33.0	24.5	42.0	100

(Table 4) represents data for the Bani Mansour region

6-3: Al-Dair area data

The frequency of hemoglobin abnormalities in the Al-Dair area is seen in Table (5). It is obvious that variations at the level of 0,01% are quite important. Patients with sickle-cell-anemia received the greatest percentage (68.1%) in 2021, and the lowest percentage (10.6%) in 2020. The incidence of sickle cell anemia peaked in 2021 at 50.0%, while it peaked at its lowest point in 2020 and 2019 at 25.0%. Anemia type D was most prevalent in 2021, with 2019 and 2020 being the only years with no occurrences. Additionally, the highest rate of 100% for type C anemia was observed in 2020; No instances were noted in 2019 or 2020. The majority of thalassemia-major-patients was recorded in 2021, reaching 56.0%, and the lowest rate in 2020, which was 20.0%. The highest rate of patients with thalassemia minor was recorded in 2021, amounting to 69,6%, and the lowest rate was 12.5% in 2020.

Disease	2019	2020	2021	Total
Sickle Cell Anemia Carrier AS	21.3	10.6	68.1	100
Sickle Cell Anemia SS	25.0	25.0	50.0	100
Type D Anemia Carrier	0.0	0.0	100.0	100
Type C Anemia Carrier	0.0	100.0	0.0	100
Thalassemia Major THM	24.0	20.0	56.0	100
Thalassemia Minor THM	17.9	12.5	69.6	100

(Table 5) represents the data of Al-Dair district

7-3: Different Incidence Rates Between The Sexes

Table (6) shows the difference in the infection rates between the sexes, and it is clear that there are significant differences, as it gave the highest value of 50.5 for females with sickle cell anemia and the highest value of 56.7 for sickle cell anemia for males, as well as giving the highest value for anemia type D for males with a rate of 77.8%, while the highest was given The value for females of anemia type C was 66.7%, while the highest value was given for patients and carriers of beta thalassemia for females, with rates of 54.5% for the infected and 55.5% for the carriers.

disease sex	Tm	TM	C	D	SS	AS	%
male	**44.5**	**45.5**	**33.3**	**77.8**	**56.7**	**49.5**	**47.2%**
female	**55.5**	**54.5**	**66.7**	**22.2**	**43.3**	**50.5**	**52.8%**
Total	**100**	**100**	**100**	**100**	**100**	**100**	**100**
P valua	**0.04 and a chi-square value of 11.125**						

(Table 6) represents the difference in infection rates between the sexes

8-3: Difference In Infection Rates Among Different Age Groups

Table (7) The difference in infection rates between the different age groups. There are highly significant differences, as the highest rate reached 20.0% for patients with sickle cell anemia and patients with thalassemia major in the age groups (1-10), while the highest rate reached 55.7% for carriers of sickle cell anemia in the other groups. The age group (11-21) and the lowest rate was 22.2% for patients with anemia type C. While the highest percentage of patients with anemia type C was in the age groups (22-32) and reached 66.7, and the lowest percentage of sickle cell anemia reached 26.4%. The highest percentage was 11.1% for patients with anemia type C in the age groups (33-43), and the lowest percentage was 5.0% for carriers of sickle cell anemia, while no case of anemia type D was recorded. The highest percentage was recorded for patients with thalassemia minor, the lowest percentage for thalassemia major, and no cases were recorded for the rest of the diseases within the age groups (54-44) and (55-65). The highest percentage was 0.6 for patients with thalassemia major, and the lowest percentage was 0.2% for patients with thalassemia major within the age groups (66-76). It is clear that the highest percentage of all diseases was 46.6% in the age group (11-21) and the lowest percentage was 0.2% among the age groups (66-76).

disease age	Tm	TM	C	D	SS	AS	%
(1-10)	9.6	20.0	0.0	11.1	20.0	9.9	12.8
(11-21)	43.5	40.3	22.2	44.4	36.7	55.7	46.6
(22-32)	31.7	30.1	66.7	38.9	36.7	26.4	29.9
(33-43)	8.4	5.5	11.1	0.0	6.7	5.0	6.3
(44-54)	4.3	2.3	0.0	5.6	0.0	2.5	3.0
(55-65)	2.2	1.2	0.0	0.0	0.0	0.0	1.0
(66-76)	0.2	0.6	0.0	0.0	0.0	0.5	0.2
Total	100	100	100	100	100	100	100

(Table 7) The difference in infection rates among different age groups

DISCUSSION.

The results of our current study showed, according to the data collected, that hemoglobin disorders constituted 8.34% of the total number of patients who performed the HPLC test to detect hemoglobin disorders. The prevalence of hemoglobin disorders increased from 30.7% in 2019 to 46.3% in 2021. The trait of beta thalassemia constituted the highest percentage of the abnormal hemoglobin group, amounting to 60.6% (761), sickle cell disease formed the percentage of (466) 37.1%, while the percentage of anemia type C, D (9-18) reached (0.8, 1.5%) on respectively.

A study conducted by (Sarkar et al., 2022) showed results similar to our current study, where beta-thalassemia was the dominant hemoglobin disorder, amounting to 758 cases (78.71%) out of 963 cases at the end of 2020.References

The percentage of cases distribution by gender was 47.2% for males and 52.8 for females. The age group that recorded the highest number of cases ranged between (11-21) years, with a rate of 46.6%. The second and third age groups were (22-32) years, with a rate of 29.9% and (29.9%). 1-10) years at a rate of 12.8%. This is contrary to what was stated in the study conducted in Kurdistan in northern Iraq, where the rates of distribution of cases by sex reached 48.75% for females and 51.25 for males. As for the age groups that reached the highest percentage of cases, they ranged between (6-15) years, with a rate of 44.45%. As for the second and third age groups, they were (1-5) years at a rate of 23.20%, and (16-25) years at a rate of 21.60% (Sarkar et al., 2022).

This may be due to the fact that the Arab countries represent a wide area that includes North Africa, the Nile Valley, West Asia and the Arabian Peninsula, in addition to the geographical differences, each country witnessed mixing from different population groups throughout history, moreover, migration between Arab countries was common until the time Present, the heterogeneity of Arab peoples was reflected in the 52 mutations detected in thalassemia, and these mutations were of Mediterranean and Asian origin, although some countries have unique mutations, there does not seem to be a specific mutation that is restricted to Arabs (Zahed, 2001; Akhavan-Niaki et al., 2011).

The frequency of thalassemia, especially -thalassemia, is rather high in Iraq, and various studies have found that the population of that country has a variety of thalassemia mutations (Al-Allawi et al., 2013; Al-Allawi and Jubrael, 2014; Musa et al. al., 2017). Or In Iraq, it could be due to the high level of consanguineous marriage, which increases the possibility of inherited genetic disorders (Hamamy and Al-Allawi, 2013).

The statistics gathered show that the prevalence of hemoglobinopathies grew from 2019 to the end of 2021 at a pace of 30,7%, reaching 46,3%. The most prevalent hemoglobinopathy in the current study was thalassemia syndrome, particularly thalassemia major, which is consistent with a study by(Sarkar et al,.2022). This rise may be attributed to the high number of consanguineous marriages and the lack of effective prevention programmes, legislation and there is an emerging demand for a preventive programme, necessitating identification of carriers, provision of genetic counseling, guidelines for distinguishing between minor and thalassemia-specific anemias, prenatal diagnosis, public education and sustainable legislation. Sarkar et al., (2022). (Kurdistan research is the source).

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