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A demographic study of sickle cell anemia patients in Al-Diwaniyah and Al-Najaf governorates

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***Abstract*---**Background: Sickle cell anemia is a common genetic disease in the world that results from a genetic mutation in a single amino acid base in the beta chain involved in the composition of hemoglobin, where the appearance of sickle hemoglobin Hbs as a result of replacement (β6Glu>Val) leads to changes in the structure of normal hemoglobin HbA. Aim of the study: Is to evaluate the demographic parameters levels in sickle cell anemia condition of patients with sickle cell anemia in the governorates Al-Diwaniyah and Al-Najaf Al-Ashraf. Methods: Patients with sickle cell anemia (n=84) 48 males and 36 females were on follow up in the Al-Diwaniyah and Al-Najaf Center for Hereditary Blood Disease, Iraq. The healthy persons (n=40) 20 males and 20 females as controls. Conclusion: The results showed that was no significant difference in the frequency distribution of patients and control subjects according to gender, a significant difference in the frequency distribution of patients and control subjects according to age, a significant difference in the frequency distribution of patients and control subjects according to body weight, and a significant difference in the frequency distribution of patients and control subjects according to parent consanguinity.

***Keywords*---**demographic study, sickle cell, anemia patients.

Introduction

Sickle Cell Anemia (SCA) with parental consanguinity found that hereditary blood diseases such sickle cell anemia is significantly more common in communities with high consanguinity marriage rates (AI Arrayed, 2005; Memish and Saeedi,

2011). The prevalence and incidence of autosomal recessive illnesses, as well as the infant mortality and morbidity rates, are the primary genetic consequences of cousin marriages (DeBaun, 2014). First cousin marriage rates in Arab nations are significantly higher than those of South and North Americans, Europeans, South Africans, eastern Asians, and the populations of Oceanic nations. These rates, which range from 40% to 50%, have been linked to social and traditional practices meant to preserve property within families (Yawn *et al.,* 2014). Patients with non-consanguineous parents had greater HbF levels than those with consanguineous parents (Al-Ghazaly *et al.*, 2013; Kahhaleh *et al.,* 2019).

A wide range of clinical manifestations resulted in considerably lower mean body weight and BMI in SCD individuals (Helvaci & Kaya., 2011). Approximately 70 genes may play a role in the regulation of bone mass, and certain genes have been demonstrated to impact both the BMI and bone geometric characteristics (Xu *et al.*, 2005). Substantial additive genetic control of the Brahman body weight to hip height ratio has also been observed (Riley *et al.*, 2007). Leptin is an adipose hormone mostly generated by adipocytes that regulates body weight centrally (Considine *et al.,* 1996). It is a skeletal growth factor that is also expressed on osteoblasts and stimulates bone mineralization (Reseland *et al.,* 2001).

Children are more likely to have strokes than adults are to have hemorrhagic strokes. Patients with SS who were younger than 20 years old had a higher incidence of infarctive stroke than those who were older, while patients over 30 years old were equally at risk. It's interesting to note that children under 10 years of age had a greater rate of hemorrhagic stroke during the era of lowest risk for infarctive stroke (20 to 29 years of age) (Ohene et al., 1998). Female gender was a significant independent predictor of neuropathic pain medication usage compared to male gender, with female patients 1.5 times more likely to receive a neuropathic medication prescription. In terms of general pain issues, women report more acute and chronic pain disorders and experience higher levels of pain-related suffering such fibromyalgia, arthritis and headaches (Lempp *et al.*, 2009).

Methods

Subjects

The number of individuals targeted by this study was (84) individuals, who were already diagnosed as carriers of sickle cell anemia by the medical advisory staff according to the clinical examination, symptoms, chromatography and blood film, who were under Follow-up in centers of Genetic blood diseases, Thalassemia patients in Al-Diwaniyah city and Al-Najaf Al-Ashraf city. The samples of the study included (48) male and (36) female and aged between (2 - 50) years and the patients individuals were divided into three groups. (40) Healthy people were randomly selected from primary health care centers that served as the control group for the study and the control group was the same age and sex for the patients.

Blood Samples

The current study period extended from the beginning of November 2021 until the end of March 2022, Blood samples were obtained from venipuncture of patients and control using a single-use syringe (5 ml). Then the blood samples were divided into groups that were 2 and 3 mL, the first group was placed in sterile tubes of tube that prevent blood clots and used for analysis of blood parameters including CBC and genetic detection, the second group was placed in normal tubes and expelled at a speed of (3000) rpm for (10) minutes to get the serum and keep it in the freezer (-20C°) until use unless used immediately to analyze biochemical parameters such as Ferritin and Creatinine and Urea.

Study area

The samples was collected from Al-Diwaniyah Governorate, It is one of the governorates of the Middle Euphrates region. Location of the province is determined between latitudes 31.17 and 32.24, and longitudes 44.24 and 45.49 east, And Al-Najaf Governorate is one of the provinces in central Iraq, located on the edge of the western plateau of Iraq, southwest of the capital, Baghdad, Longitude 32.0259, Latitude 44.3462.

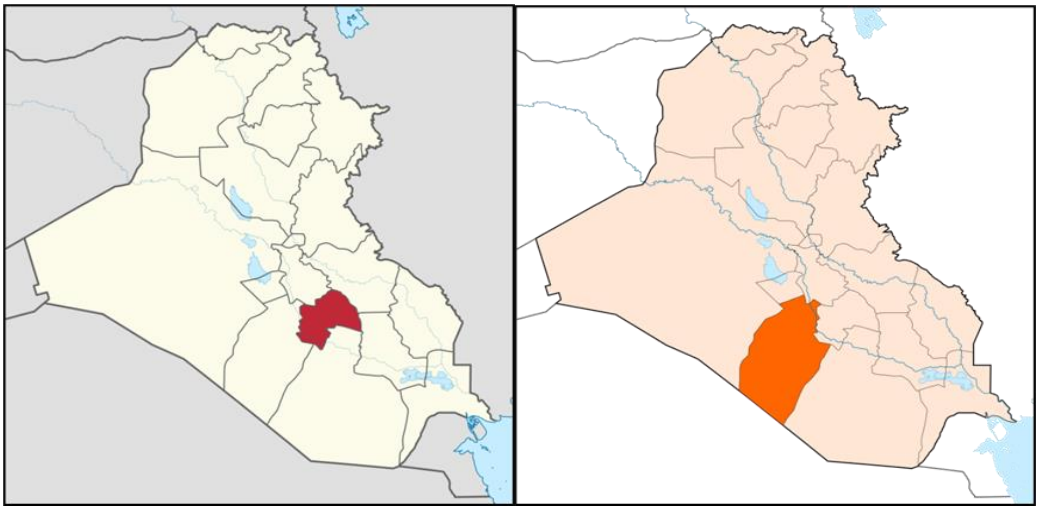

Fig (1-1): Study area

Results

The present study enrolled 84 patients with sickle cell anemia and 40 apparently healthy subjects. The demographic characteristics of patients and control subjects are shown in table (1) there is no significant difference in the frequency distribution of patients and control subjects according to gender, Patients' group included 48 (57.14 %) males and 36 (42.85 %) females, whereas, control group included 20 (50.0 %) males and 20 (50.0 %) females and there was no significant difference in the frequency distribution of patients and control subjects according to gender (P = 0.602) Fig (1-1).

The table (1) showed that a significant difference in the frequency distribution of patients and control subjects according to age, Patients' group included 3-18 years 39 (46.42 %), whereas, control group included 10 (25.0 %), 19-34 years 25 (29.76 %), whereas, control group included 16 (40.0 %) and 35-51 years 20 (23.80 %), whereas, control group included 14 (35.00 %), and there was a significant difference in the frequency distribution of patients and control subjects according to gender (P = 0.006) Fig (1-2).

The table (1) reported that no significant difference in the frequency distribution of patients and control subjects according to blood groups, Patients' group included A^+, 24 (28.57 %), whereas, control group included 10 (25.0 %), A^-, 2 (2.38 %), whereas, control group included 4 (10.00 %), B^+, 24 (28.57 %), whereas, control group included 8 (20.0 %), B^-, 0 (0.00 %), whereas, control group included 3 (7.5 %), AB^+, 2 (2.38 %), whereas, control group included 5 (12.5 %), O^+, 32 (38.09 %), whereas, control group included 9 (22.5 %) and O^-, 0 (0.00%), whereas, control group included 1 (4.0 %), there was no significant difference in the frequency distribution of patients and control subjects according to gender (P = 0.071) Fig (1-3).

The table (1) shows a significant difference in the frequency distribution of patients and control subjects according to body weight, Patients' group included 14-37 kg 10 (11.90 %), whereas, control group included 0 (0.00%), 38-61 kg 54 (64.28 %), whereas, control group included 10 (25.0 %) and 62-85 kg 20 (23.80 %), whereas, control group included 30 (75.0 %), and there was a significant difference in the frequency distribution of patients and control subjects according to gender (P = 0.00) Fig (1-4).

The table (1) reported a significant difference in the frequency distribution of patients and control subjects according to parent consanguinity, Patients' group included yes 72 (85.71 %), whereas, control group included 15 (37.5 %), and no 12 (14.28 %), whereas, control group included 25 (62.5 %), and there was a significant difference in the frequency distribution of patients and control subjects according to gender (P = 0.00) Fig (1-5). The table (1) demonstrated a significant difference in the frequency distribution of patients and control subjects according to spleen status, Patients' group included present 43 (51.19 %), whereas, control group included 40 (100.00 %), and splenectomy 41 (48.80 %), whereas, control group included 0 (0.00 %), and there was a significant difference in the frequency distribution of patients and control subjects according to gender (P = 0.00) Fig (1-6).

Table (1) Demographic characteristics of study groups

Characteristic	Category	Groups		X^2/P-value
		Control N=40	Patients N=84	
Gender	Male	20(50.0)	48(57.14)	0.272/0.602
	Female	20(50.0)	36(42.85)	
Age	3-18	10(25.0)	39(46.42)	10.26/0.006*
	19-34	16(40.0)	25(29.76)	
	35-51	14(35.0)	20(23.80)	

Blood groups	A+	10(25.0)	24(28.57)	11.62/0.071
	A-	4(10.0)	2(2.38)	
	B+	8(20.0)	24(28.57)	
	B-	3(7.5)	0(0)	
	AB+	5(12.5)	2(2.38)	
	O+	9(22.5)	32(38.09)	
	O-	1(4.0)	0(0)	
Body weight	14-37	0(0)	10(11.90)	22.34/0*
	38-61	10(25.0)	54(64.28)	
	62-85	30(75.0)	20(23.80)	
Parent consanguinity	Yes	15(37.5)	72(85.71)	30.73/0*
	No	25(62.5)	12(14.28)	
Spleen status	Present	40(100.0)	43(51.19)	25.84/0*
	Splenectomy	0(0.0)	41(48.80)	

* Significantly difference at P<0.05

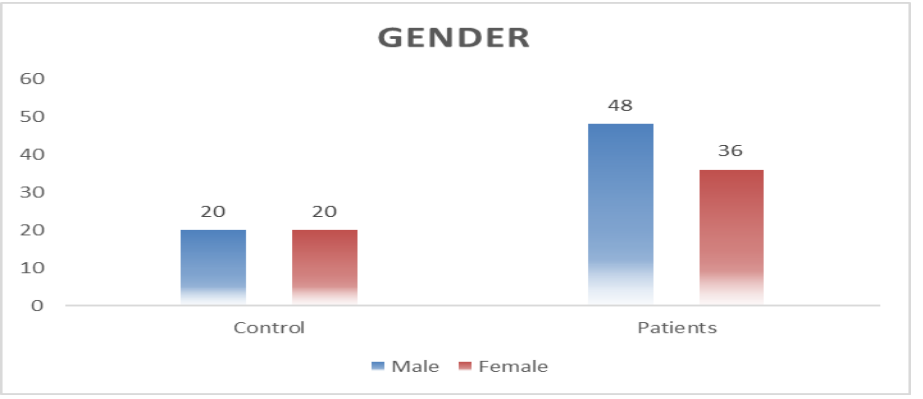

Fig (1 – 2): Distribution of patients and control group according to gender

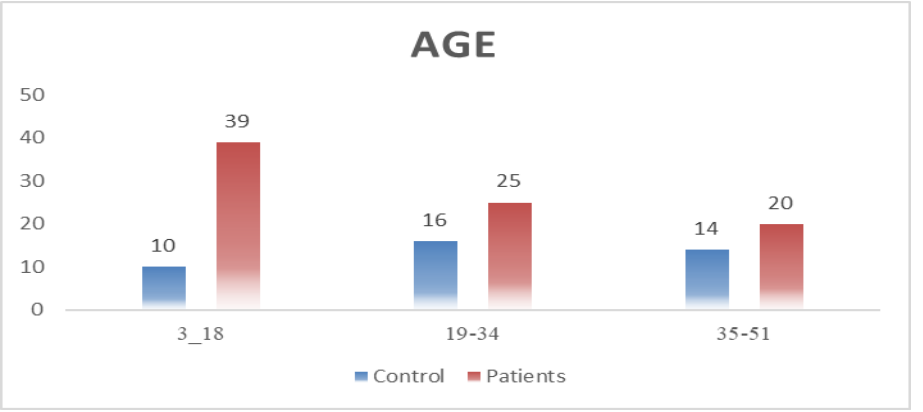

Fig (1 – 3): Distribution of patients and control group according to age

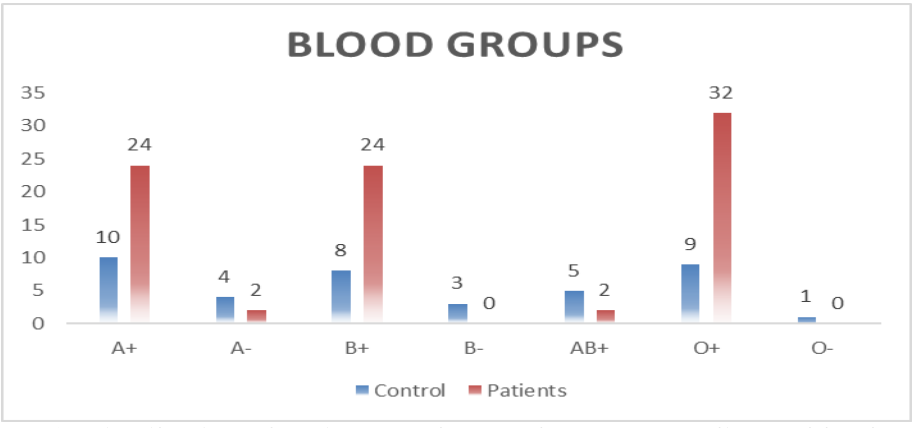

Fig (1 – 4): Distribution of patients and control group according to blood groups

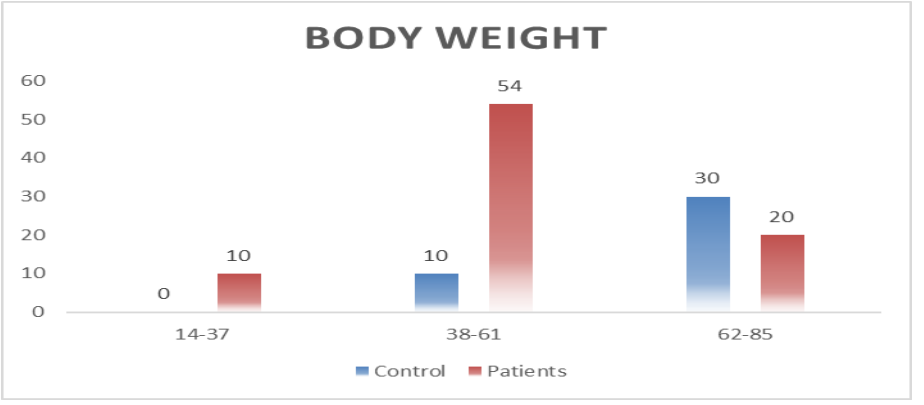

Fig (1 – 5): Distribution of patients and control group according to body weight

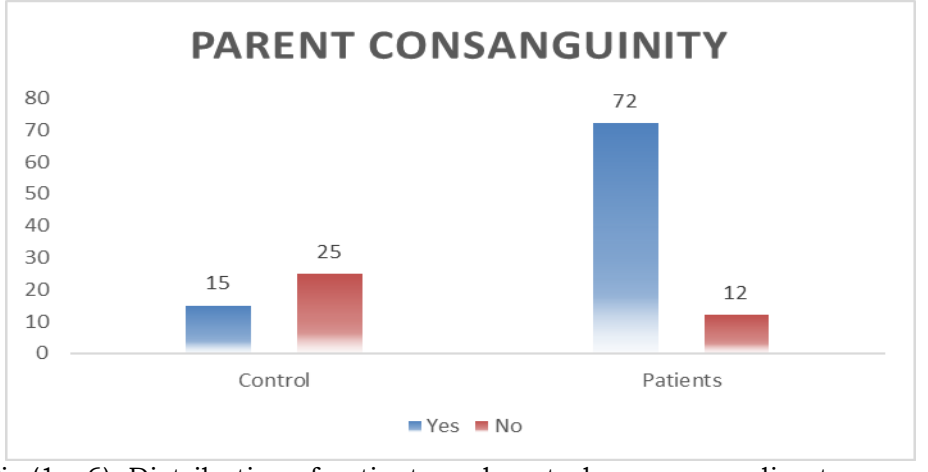

Fig (1 – 6): Distribution of patients and control group according to parent consanguinity

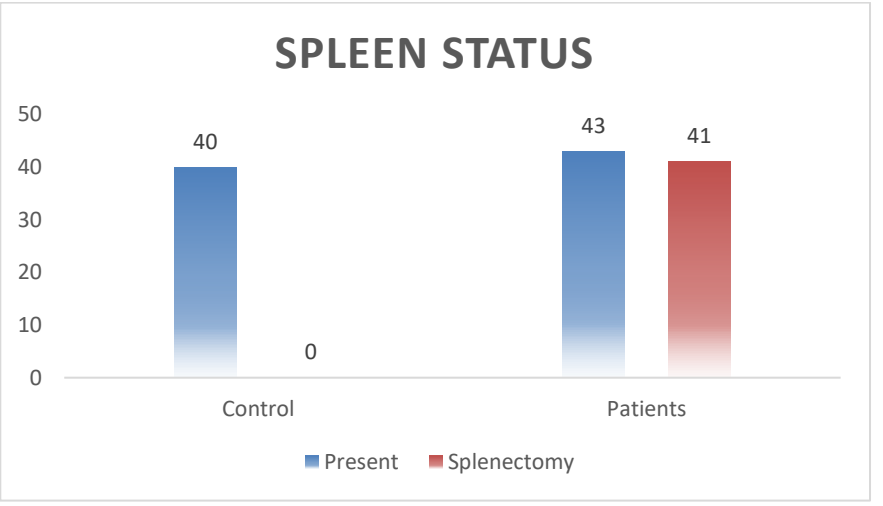

Fig (1 – 7): Distribution of patients and control group according to spleen status

Discussion

The current study recorded a significant difference in the frequency distribution of patients and control subjects according to parent consanguinity in sickle cell anemia patients compared with control group, that's agree with the results of the study of Al-Ghazaly *et al.* (2013) and Kahhaleh *et al.* (2019), who reported patients with non-consanguineous parents had greater HbF levels than those with consanguineous parents. At birth weight in both boys and girls were similar to healthy, but fell away thereafter. By age 18 years, female patients had caught up with healthy but male patients, many of whom were still growing, had not yet caught up (Thomas *et al.,*2000). Splenomegaly was present in SCA patients older than 10 years old. RBC and Hb mean values were considerably lower in children with SCA, although MCHC, WBC, platelet, and reticulocyte mean values were significantly greater (Abdullahi *et al.,* 2014).

The present study recorded a significant difference in the frequency distribution of patients and control subjects according to spleen status in parent consanguinity in sickle cell anemia patients compared with control group, That agree with the results of the study of Owusu-Ofori and Remmington (2017) who reported that SCA patients suffer from the deposition of red blood cells in the spleen, which leads to painful spleen isolation crises. The spleen is removed completely or partially to reduce the risks of these crises and stop blood transfusions, but this is not without the risks of infection and blood poisoning in patients and children, especially and the loss of the immune protection it provides spleen for body.

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