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Sickle Cell Disease in the Zinder Region in 2023: Prevalence and Sociodemographic Factors

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Abstract

Introduction: In view of the number of sickle cell patients and due to a low production of descriptive studies, we decided to determine the prevalence of genes S and C of the disease in the Zinder region. The objective was to contribute to improving the management of sickle cell disease in Zinder. **Methodology:** This was a systematic screening by the "Sickle Scan" test of any blood donor admitted to the Zinder Regional Blood Transfusion Center during the 6-month study period, from January to June 2023. The Sickle Scan is a qualitative lateral flow chromatography immunoassay using whole blood samples that aid in the rapid diagnosis of sickle cell disease. **Results:** The study was carried out on 613 samples during the period concerned. The frequency of sickle cell genes was 26.9% (n = 165) in all samples collected, with 23.1% (n = 142) and 3.8% (n = 23) for the S gene and the C gene, respectively. The 18 - 30 age group was the most represented with 64.4% (n = 395) cases. The median age of blood donors was 26 years ± 10 years (min = 18 years/max = 60 years). The sex ratio was 2.5. Donors of Nigerien nationality accounted for 84.1% (n = 516). There is a predominance of blood donors with an average monthly income between 34,000 and 70,000 CFA francs in 44.3% (n = 272), lived in permanent housing with drinking water supply. Sickle cell trait (SMA) was found in 22.5% (n = 138). **Conclusion:** The analysis of these results highlights a high frequency of the S gene for sickle cell disease. The population with an average monthly income is the most affected, with a male predominance.

Keywords

Sickle Cell Anemia, Blood Donors, Sickle Scan

1. Introduction

Sickle cell disease, also known as sickle cell anemia, is a genetic hemoglobin disorder with autosomal recessive inheritance [1]. It results from a single point mutation in the sixth codon of the ß-globin gene (GAG → GTG: replacement of glutamic acid by valine in the sixth position on the ß chain of globin), leading to the production of abnormal hemoglobin (HbS). The polymerization of deoxygenated HbS is responsible for chronic hemolytic anemia and vaso-occlusive events [1]. Sickle cell disease predominantly affects black populations, but it is not exclusive to them, as cases have been reported in authentic Caucasians, Arab populations, etc [1]. An area known as the "sickle belt" has been described, stretching across Africa from south of the Sahara to north of the Zambezi, between the 15th parallel north and the 20th parallel south [1]. According to WHO, nearly 5% of the global population carries a gene responsible for abnormal hemoglobin [2]. The prevalence of the sickle cell gene in Africa ranges from 10% to 40%, with rates of 5% to 20% in West Africa and 8% to 24% in Niger [1]. It is a serious lifelong disease that affects millions of people in Africa, according to the WHO. Its prevalence is increasing due to a lack of awareness and care infrastructure in Zinder. In the course of this study, we intend to strengthen the prevention of the transmission of the disease gene and reduce the prevalence of sickle cell disease in the Zinder region. In view of the number of patients and the public health problem of sickle cell anemia in Niger, we initiated this study on sickle cell anemia in the Zinder region. Through this study, we aim to strengthen the prevention of gene transmission and reduce the prevalence of sickle cell disease in the Zinder region. The objective of this study is to contribute to improving the management of sickle cell disease in Zinder.

2. Patient and Methods

This was a cross-sectional study with prospective data collection conducted over a period of 6 months, from January to June 2023. The study included all blood donors admitted for voluntary or family donation at the regional blood transfusion center in Zinder. In our study, we included asymptomatic voluntary or family blood donors who agreed to undergo the Sickle Scan test. The Sickle Scan test is a confirmatory test that detects hemoglobins S, C, and A. It is a qualitative immunological test using lateral flow chromatography, performed with a sample of whole blood, which aids in the rapid diagnosis of sickle cell disease. The Specificity and sensitivity of the Sickle SCAN Rapid Test was evaluated by comparison with hemoglobin electrophoresis, one of the standard lab tests for sickle cell diagnosis. The Sickle SCAN® has both a combined sensitivity and specificity of > 99% and provides results in 5 minutes, making it one of the fastest and most accurate tests in the world (BioMedomics Sickle SCAN, tests for life). We conducted a comprehensive recruitment of all donors meeting our inclusion criteria during the study period. In total, 613 patients were included. The measured variables included the frequency, which determined the proportions of sickle cell

disease genes (S and C) among all analyzed samples, sociodemographic characteristics (age, sex, socioeconomic level, place of origin), and diagnostic variables (types of hemoglobin, distribution of hemoglobin types according to age, sex, region of origin, and monthly income). Data collection and statistical analyses were performed using Epi Info 7.2.2.2. Our results were presented in the form of tables and figures. Microsoft Office 2016 Word and Excel software were used for data entry and designing tables and figures. The results are expressed as mean ± standard deviation and as percentages of individuals. Central tendency values (mean, minimum, maximum, median) were determined for certain quantitative variables.

3 Results

Of the 613 tests performed, there was a prevalence of 26.9% (n = 165) of hemoglobin S and hemoglobin C of sickle cell disease. The genes for abnormal haemoglobin S and C were found in 23.16% and 3.74% of cases, respectively. The 18 to 30 age group predominates with 64.44% (n = 395). The median age of patients was 26 years ± 10 years (min = 18 years/max = 60 years). There is a predominance of the male sex with a sex ratio of 2.54. The Nigerien population predominates with 516 cases, or 84.17%. Blood donors with an average monthly income of between 34,000 and 70,000 CFA francs were in the majority in 44.40% of cases. The socio-demographic aspects are summarized in Table 1.

Table 1. Distribution of patients by sociodemographic aspects.

Variables		n (%)
Age/year	[18 - 30]	395 (64.44)
	[30 - 40]	105 (17.13)
	[40 - 50]	67 (10.93)
	[50 - 60]	38 (6.20)
	≥60	8 (1.31)
Sex	Feminine	173 (28)
	Masculine	440 (72)
Country of origin	Niger	516 (84.17)
	Togo	3 (0.50)
	Tchad	35 (5.71)
	Cameroun	7 (1.14)
	Bénin	51 (8.32)
	Nigeria	1 (0.16)
Monthly income	<34,000	212(34.60)
	[34,000 à 70,000]	272 (44.40)
	>70,000	129 (21.00)

There is a predominance of blood donors with normal hemoglobin AA_2 in 73.10% of cases. In 26.9% (n = 165) of cases, the presence of the sickle cell gene S or C was found in blood donors at the Zinder Regional Transfusion Centre. Sickle cell trait (AS) and (CA) are distinguished in 22.51% (n = 138) and 3.42% (n = 21) of cases, respectively. Asymptomatic major sickle cell syndromes (MDS) were found in our study and included homozygous sickle cell anemia (SS) in 0.65% of cases (n = 4) and composite heterozygosity (SC) in 0.32% of cases (n = 2). There was a predominance of blood donors with normal haemoglobin AA_2 in the age group of [18 - 30] in 48.77% (n = 299/613). There is a predominance of abnormal haemoglobin in blood donors from Niger, Benin and Chad in 23.16% (142/613); 3.26% (20/613); 0.48% (3/613) of cases respectively. There is a predominance of male sex with normal hemoglobin AA_2 in 52.52% (322/613). There is a predominance of blood donors with sickle cell disease S or C gene in the low- to middle-income population in (n = 125/165) 20.39% of cases.

The poor population is the most affected by the disease. There is a predominance of abnormal haemoglobin in blood donors from Niger, Benin and Chad in 23.16% (142/613); 3.26% (20/613); 0.48% (3/613) of cases respectively. The clinical aspects are summarized in Table 2.

Table 2. Distribution of patients by clinical aspects.

Variables		n (%)				
		AA	AS	AC	SS	CC
Sickle Scan result		448 (73.10)	138 (22.51)	21 (3.42)	4 (0.65)	2 (0.32)
Type of haemoglobin according to age groups	[18 - 30]	299 (48.77)	76 (12.39)	17 (2.77)	1 (0.16)	2 (0.32)
	[30 - 40]	75 (12.23)	28 (4.56)	2 (0.32)	0 (0.00)	0 (0.00)
	[40 - 50]	39 (6.36)	24 (3.91)	2 (0.32)	2 (0.32)	0 (0.00)
	[50 - 60]	27 (4.40)	10 (1.63)	0 (0.00)	1 (0.16)	0 (0.00)
	≥60	8 (1.30)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)
Type of hemoglobin by sex	Féminine	126 (20.55)	38 (6.19)	9 (1.46)	0 (0.00)	0 (0.00)
	masculine	322 (52.52)	100 (16.31)	12 (1.95)	4 (0.65)	2 (0.32)
Type of hemoglobin by region of origin	Niger	374 (61.01)	126 (20.55)	12 (1.95)	4 (0.65)	0 (0.00)
	Bénin	31 (5.05)	9 (1.46)	9 (1.46)	0 (0.00)	2 (0.32)
	Cameroun	7 (1.14)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)
	Nigeria	1 (0.16)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)
	Tchad	32 (5.22)	3 (0.48)	0 (0.00)	0 (0.00)	0 (0.00)
	Togo	3 (0.48)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)
Hemoglobin type by monthly income	<34,000	149 (24.30)	56 (9.13)	6 (0.97)	1 (0.16)	0 (0.00)
	[34,000 - 70,000]	210 (34.25)	48 (7.83)	11 (1.79)	1 (0.16)	2 (0.32)
	>70,000	89 (14.51)	34 (5.54)	4 (0.65)	2 (0.32)	0 (0.00)

4. Discussion

During the study period, a total of 613 blood donors were tested. There is a predominance of blood donors with normal hemoglobin type AA_2 in 73.10% of cases. In 26.9% (n = 165) of cases, the presence of sickle cell trait (gene S or C) was found among blood donors at the Zinder Regional Transfusion Center. Sickle cell trait (AS) and (AC) were identified in 22.51% (n = 138) and 3.42% (n = 21) of cases, respectively. Our results are consistent with those of Jean-Benoît Arlet, who found in his study on the epidemiology of sickle cell disease in France and worldwide that several sub-Saharan African countries exceed the 20% incidence of gene carriers at birth [3].

We observed a predominance of abnormal hemoglobin among blood donors from Niger, Benin, and Chad in 23.16% (142/613); 3.26% (20/613); and 0.48% (3/613) of cases, respectively. In Senegal, 1 out of 10 individuals, regardless of ethnicity, geographical origin, or social class, carries the sickle cell gene. These are mainly individuals with sickle cell trait (AS) [4]. The majority of people affected by this disease live in sub-Saharan Africa, with prevalence rates ranging from 10% to 40% **[5]**. Two African countries, Nigeria and the Democratic Republic of Congo, have the highest prevalence due to their large population. India is another country with a high number of births of sickle cell patients [6]. In fact, 45% of sickle cell adults followed in the National Reference Center of the European Hospital Georges-Pompidou were born in Africa (mostly sub-Saharan) [7]. In the United States, heterozygotes represented 8% to 13% of individuals of African descent in a study published by the University School of Medicine [8]. Sickle cell disease is predominantly a disease of the black population that dramatically affects Africa.

We observed a predominance of individuals with sickle cell trait (gene S or C) in the low to intermediate-income population in 20.39% (n = 125/165) of cases. Our results confirm the work of the English geographer Frédéric Piel who, over the past twenty years, has provided important epidemiological data on the estimation of births in low-income countries and their foreseeable evolution for the next thirty years [9]. The underprivileged population is most affected by sickle cell disease. Abnormal hemoglobin was found in 15.66% (96/613) of cases in the age group [18 – 30]. In the age group [50 – 60], the rate of abnormal hemoglobin was 1.79% (11/613) compared to 4.40% (27/613) for normal hemoglobin.

A recent study conducted in five francophone African countries highlights a lower mortality rate among sickle cell patients compared to previous estimates [10] [11]. This indicates an improvement in the quality of life for individuals with sickle cell disease. Optimal management, as carried out in referral centers, along with the introduction of new drugs to treat abnormal hemoglobins [12] [13] [14], is transforming the lives of patients. Many are reaching adulthood, receiving education, having a profession, and starting families.

In our study, males represented more than half of the subjects with abnormal hemoglobin (118/47) with a ratio of 2.51. This male predominance was also

found in Boiro's study, which reported a sex ratio of 1.42 **[15]**.

However, other authors such as Nacoulma and Samira reported a female predominance [16] [17]. Finally, other authors found no predominance between the two sexes, such as Thuilliez [18] and Dreux [19]. These differences may be related to the demographic data of each country since the transmission of sickle cell disease is not linked to gender [20] [21], and blood donation is more common among men than women in our context.

5. Conclusion

The sickle cell gene is common in the Zinder region, as evidenced by the prevalence obtained during our study. It is an abnormality of red blood cell hemoglobin that affects a significant portion of the population in Zinder. The manifestation of these abnormal hemoglobin genes in double alleles leads to sickle cell disease. As the most common genetic disease worldwide, it is recognized as a debilitating condition. Sickle cell disease primarily affects underprivileged populations in our developing countries. Optimal management transforms the lives of patients. This involves strengthening the prevention of gene transmission for the disease. Mastering these epidemiological factors is necessary to achieve such a result.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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