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Sickle Cell Trait and Blood Groups (ABO and Rh) in Angolans Submitted to Hemoglobin Electrophoresis

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Keywords: Sickle cell trait; Blood group; Hemoglobin electrophoresis; Angolan population.

Abstract

Background: The sickle cell anemia is a chronic disease, incurable, although treatable, the number of cases of sickle cell anemia in Angola is considered worrying, in Luanda and it is estimated that around 16,000 babies are born with anemia in Angola each year sickle cell disease.

Objective: Estimate the sickle cell trait and its relationship with blood groups (ABO and Rh) among Angolan users who perform hemoglobin electrophoresis in the Mediag laboratory.

Methodology: The study was conducted as a cross-sectional study and quantitative approach.

Results: Of the 59 participants, the incidence of sickle cell trait in 64% (38/59), of these 22% (13/59) had homozygosis (HbSS)and 42% (25/59) had heterozygote (Hb AS), 92% (12/13) of sickle cell anemia participants were younger and the average age of participants with sickle cell disease was 11 years (SD=7.4), Rh(+) participants with sickle cell disease was 84% (11/13), with sickle cell trait was 80% (20/25), the blood group O-Rh(+) was the most affected with 45% (6/13) of participantsHb SS, the blood groups A and O for Rh(+) together accounted 60% (15/25) of sickle cell trait participantsHb AS, 60% (8/13) of homozygotes participants(Hb SS) for the sickle cell were treated with Hydroxyurea and Persantin, 44% (26/59) of participants had a direct family history of sickle cell disease and in this group 58% (15/26) has sickle cell trait heterozygote (HbAS) and 35% (9/26) has sickle cellhomozygote (HbSS), 41/59 (69.5%) of participants do not know what sickle cell anemia is, especially participants without sickle cell trait (Hb AA) and with sickle cell trait (Hb AS).

Conclusion: The incidence of sickle cell trait was high among study participants and although the number of participants is small to estimate the incidence of sickle cell anemia in the Angolan population, the incidence of the disease and trait is high among individuals suspects, especially among those with a family history of the disease, the blood group O-Rh (+) and AB-Rh (+).

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Background

The sickle-cell disease is an autosomal recessive blood disorder characterized by abnormal red blood cells due to a mutation in the haemoglobin-encoding gene. It is also known as black race disease because of its origin. Historically, the highest frequency of haemoglobin variant was found in Africa, mainly in the Midwest and South West-Atlantic regions. Currently, this chronic and incurable disease is found in several countries around the world due to miscegenation between different races [1-3]. In Africa, this disease is common in area such as sub-Saharan equatorial regions, located north of the Kalahari Desert where it appears been a natural barrier to the expansion of Plasmodium falciparum (malaria-transmitting mosquito). Annually more than 300 000 childs are born with sickle-cell diseases each year in Africa, the disease prevalence is 20-30% in Cameroon, Republic of Congo, Gabon, Ghana and Nigeria while in Uganda it is around 45%. The majority of these children die before 5 years old (OMS, 2020). In Luanda, approximately 16,000 babies are born with sickle cell disease each year. Annually, the David Bernardino Paediatric Hospital of Luanda receives about 1,500 new patients with this disease in spite of there is no neonatal screening [4,5].

The genetic haemoglobin mutation occurs in chromosome eleven where the sixth codon of the beta chain (β) is located. This mutation involves DNA´s two nitrogenous bases and an exchange of thymine by adenine happens in. Thymine is characterized by participation of glutamic acid that is rich in energy supply and aids in metabolism and brain functioning. In some case, the glutalamic acid is replaced by the amino acid valine, which is an essential amino acid and, consequently, it forms a haemoglobin polymerizes leading to sickle cell formation [1]. The frequency of mutant haemoglobin reaches 25% in population of regions such as Gambia and Senegal rivers, West Africa of the Atlantic coast, West of Central Africa and Congo River in Central Africa [6]. In Angolan territory, the presence of an allele of the disease is about 20% of the population leading to a natural incidence of 15 in 1000 born that have both alleles [6,7]. According to the head of the Luanda Paediatric Hospitals patient support services, Brígida Santos (2016), it is a public health problem in Angola that requires urgent fighting, because of its consequences on the population health [7].

Heterozygosis for haemoglobin S (sickle cell trait) is a relatively common and clinically benign situation. Most patients with sickle cell trait do not have adverse clinical signs and symptoms, except for some reports of clinical changes at high altitudes (above 3200 meters and in flights with depressurized cabin). From a haematological point of view, global counts and erythrocyte morphology are normal, as well as red cell survival, so these individuals do not have anaemia or haemolysis and their clinical follow-up should be performed in the same way as in the Hb AA population (normal) as they will often be exposed to the same medical problems [8]. The disease seems to be unknown in the first 3 to 6 months of life due to the higher percentage of Hb. In the patient with SS genotype, F tends to reduce the clinical manifestations of the patients by inhibition of Hb Spolymerization [9,10]. Thus, sickle cell disease is often asymptomatic in the first months of life, and as the amount of fetal haemoglobin decreases after 3 to 6 months, increases the likelihood of complications, especially bacterial infections, including pneumococcal [9]. The most common clinical manifestations include vase-occlusive painful crises, acute thoracic syndrome and bacterial infections that lead to hospital admissions and various acute and chronic complications, which are unimportant for premature morbidity and mortality, so these patients lead life expectancy significantly short [11-13].

Although treatable, this disease generally bring suffering to patients requiring attention from the medical, genetic and psychosocial is needed [5]. A study carried out in Nigeria demonstrates the protective and causal association between types of blood groups ABO and malaria as well as sickle cell anemia [13-15]. In 2014, a retrospective cohort study demonstrated that O blood group has a protective effect on vase-occlusive crisis by reducing the von Willebrand release factor [16]. This study aims to analyse the relationship between sickle cell trait and blood groups (ABO and Rh) among Angolan who perform haemoglobin electrophoresis in the Media laboratory. The results of this study can help us to know the incidence of sickle cell anemia and the sickle cell trait among individuals who receive an indication for electrophoresis examination and, with that, we will be able to promote future studies to assess the incidence of sickle cell anemia in Angola and thereby establish as a priority the inclusion of health programs that provide genetic counseling to reduce the number of people affected by sickle cell disease.

Methodology

The study was conducted as cross-sectional study and quantitative approach. The study was approved by the Human Research Ethics Committee of the Higher Institute of Health Sciences (Official Letter No. 169/GD/ISCISA/UAN/2017) and authorized by the direction of the Mediag Clinical Analysis laboratory in Luanda. All patients and/or caregivers of patients who agreed to participate in the study had to sign the informed consent form after being informed about the nature and objectives of the study.

Patient recruitment

The study population consisted of 59 of the 65 patients admitted to the Mediag laboratory for the electrophoresis examination between January and June 2017, and only those patients who met the selection criteria and agreed to participate in the study were included in the study, additional information was collected through an open and closed question questionnaire for patients aged 16 to 50 and for patients under 16 years of age the questionnaire was administered to their guardian as long as they were the patient's father or mother and residing in the same dwelling.

Diagnosis of ABO and Rh blood group andsickle cell trait

For the clinical data presented in the article, a blood sample was taken from the patients in test tubes containing EDTA (Ethylenediaminetetraacetic Acid) anticoagulant specific for the ABO and Rh blood group phenotyping tests and for the electrophoresis examination. Hemoglobin electrophoresis was performed using a device called Sebia brand Minicap, the Hb (E) minicap kit was developed to separate normal hemoglobins (A, F and A2) and to detect and quantify variant hemoglobins (including S, C, E, D).Blood group determination was performed by the microplate technique, which is an agglutination test between patient serum and Anti A, Anti B, and Anti D reagents in each of the wells for phenotypic identification of blood groups (ABO and Rh).

Statistical analysis

All information data and clinical outcome data were entered into a SPSS 20 database and analyzed for presentation of study

results and in tables, the graphs as prepared in the Sigmaplot 12 statistical program.

Results

Most participants were under 20 years old, represented almost 60% (33/59) of the participants included in the study and in this group the average age was 9 years, participants aged 20 to 40 years represented approximately 37% (22/59) of the participants followed in the study and the average age in this group was 28 years, this demonstrates that the study population was relatively young. Blood group O was found in about 41% of the total population and all participants in this blood group were under 41 years old. Of the 59 participants of the study, we obtained an incidence of sickle cell trait in 64% (38/59) of the participants studied, of these 22% (13/59) had homozygote (HbSS) and 42% (25/59) had heterozygote (Hb AS), 92% (12/13) of sickle cell anemia participants were younger than 20 years old, demonstrating the life expectancy of sickle cell anemia patients in Angola (Table 1).

Table 1: Distribution of patients by age, blood groups, gender and sickle cell trait.

AGE GROUPS	Blood Groups				Sub-total	Sickle Cell Trace			Mean Age(SD)
	A	AB	B	O	n (M/F)	AA	AS	SS	Years
<20 years	9	5	8	11	33 (11/22)	12	9	12	9,3(6,5)
20 to 40 years	5	0	4	13	22 (7/15)	8	13	1	27,9(5,3)
> 40 years	2	1	1	-	4 (3/1)	1	3	-	49,5(4,5)
Total	**17**	**6**	**13**	**24**	**59 (21/38)**	**21**	**25**	**13**	**18,9(13,5)**

(A; B; AB; O) - ABO Sanguine group phenotype. (M/F) - Male/Female. (AA; AS; SS) - genotypes for the sickle cell trait. (SD) - Standard Deviation.

It can be observed that when relating blood group results to hemoglobin electrophoresis results for sickle cell trait, only 1 of the 10 study participants Rh-negative factor had homozygous for sickle cell trait (Hb SS), these participants were from blood groups A and B, representing 15% (2/13) of partipants with homozygous sickle cell trait, however heterozygous sickle cell trait (Hb AS) was observed in 20% (5/25) of participants with Rh negative factor.The percentage of Rh-positive participants with homozygous sickle cell trait (Hb SS) was 84% (11/13) and the heterozygous sickle cell trait was 80% (20/25). Blood group O with Rh positive factor was the most affected by homozygous sickle cell trait (Hb SS), because 45%(6/13) of participants with homozygous sickle cell trait belonged to this blood group. Blood groups A and O for Rh factor positive together accounted for about 60% (15/25) of heterozygous sickle cell trait participants (Figure 1). With the exception of the group of patients with blood group AB, in all blood groups, there was a greater number of female patients than male, which was to be expected, since in the study 64% (38/59) of the participants were female (Table 1).

Although it was found that most patients with homozygous sickle cell trait were younger than 20 years old, it was found that of only 4 out of 13 patients who were homozygous for sickle cell trait (Hb SS), they did not perform any type of work or student activity and claimed that do not do so due to the frequent vaso-occlusive seizures that do not allow them to perform any activity, it was also observed that most homozygous sickle cell trait patients (8/13) were students and the complications resulting from the disease were cited as a hindrance to school success. However, the average age of most of our participants compared to those who studied and those who did not perform any activity were 6 and 15 years respectively, suggesting that most of the participants in our study were children and adolescents in school. Most of the heterozygous participants (12/25) for the heterozygous sickle cell trait (Hb AS) were found to be working individuals and this is reported to have a normal life and no clinical enhancement complication (Figure 2).

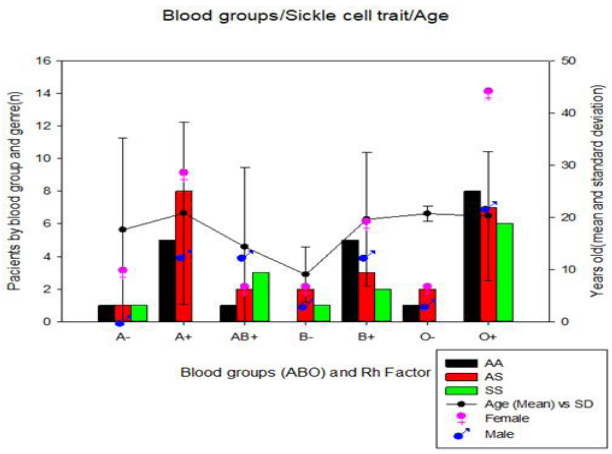

Figure 1: Blood group and Rh factor with sickle cell trait. (A): participants who show an agglutination reaction when their blood has been subjected to anti-B monoclonal antibody. **(B)**: participants who show an agglutination reaction when their blood has been subjected to anti-A monoclonal antibody. **(AB):** participants who did not show an agglutination reaction when their blood was subjected to monoclonal antibody anti-A and Anti-B. **(O):** participants who showed an agglutination reaction when their blood was subjected to monoclonal antibody anti-A and Anti-B. **(- and +):** participants who did not present and who presented agglutination reaction to the anti-D antibody, respectively.Related to the sickle cell trait, mean age and gender.

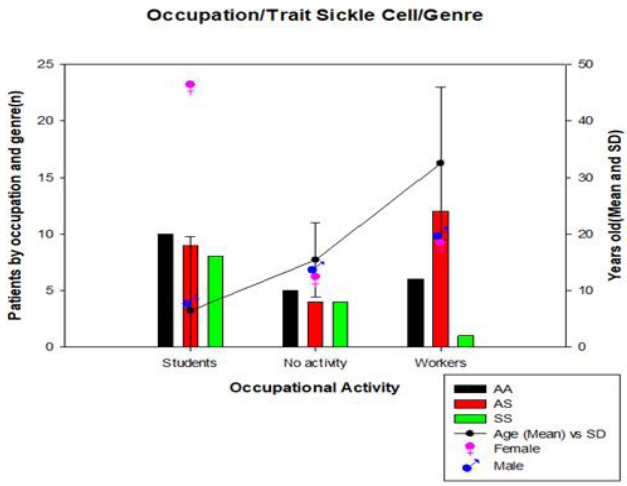

Figure 2: Sickle cell trait and occupation. **(Students)**: group of participants who said they were students and were enrolled in a school but did not carry out any other remunerative activity. **(No Activity)**: participants who mentioned in an interview that they do not carry out any type of academic or professional activity. **(Workers)**: group of participants who mentioned that they carry out some type of professional activity and receive remuneration. Related to the sickle cell trait, mean age and gender.

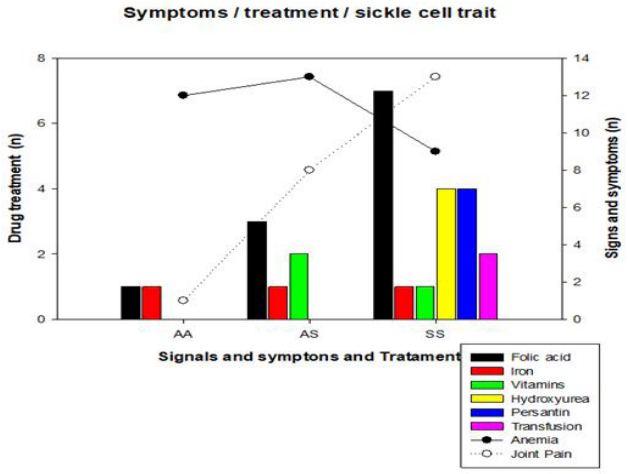

Figure 3: Patients according to symptoms and treatment. **(AA)**: group of participants who, when submitted to hemoglobin electrophoresis, the results did not present the sickle cell trait in any of the alleles (Hb AA). **(AS)**: groups of participants who, when submitted to hemoglobin electrophoresis exam, the results of the exams presented an allele with the sickle cell trait (Hb AS). **(SS)**: group of participants who, when submitted to electrophoresis examination, presented the sickle cell trait in both alleles (Hb SS).Related to signs and symptoms and treatment.

It was found in the study that all study participants claimed that the symptom that led to their consultation and need for examination was joint pain anemia. Among the homozygote for the sickle cell trait(Hb SS), joint pain was the main complaint (13/13), followed by anemia that was referred by 69% (9/13) of the participants. It was found that 30% (4/13) heterozygote for the sickle cell trait(Hb SS), even without diagnosis, were already treated with chemotherapy (hydroxyurea) and antiplatelet (Persantin), as well as 62% (8/13) were using folic acid and 15% (2/13) had already undergone blood transfusion. When compared with heterozygous participants for sickle cell trait(Hb AS), 52% (13/25) reported anemia and 32% (8/25) joint pain, but only 24% (6/25) of them were taking some kind of medication as folic acid (3/6), iron (2/6) and vitamins (1/6). Participants without sickle cell trait also reported being affected by anemia in about 57% (12/21) and joint pain in 5% (1/21), but only one reported using folic acid and the other iron (Figure 3).

In the study, 44% (26/59) of participants had a direct family history(father, mother, brother, uncle or cousin) of sickle cell disease (Hb SS) or sickle cell trait(Hb SS), and in this group of participants 58% (15/26) had sickle cell trait heterozygote (AS) and 35%(9/26) had patients with sickle cell trait homozygote (SS). Among the participants who reported not knowing about family members with the disease or sickle cell trait, only 30% (10/33) had the heterozygous sickle cell trait(Hb AS) and 15% (4/33) had the homozygous sickle cell trait(Hb SS). It was also found that among the participants without the sickle cell trait (AA) and with the hoterozygoussickle cell trait (AS), most had electrophoresis examination as a medical routine (check-up), but all participants homozygous for the sickle cell trait (SS), only underwent electrophoresis due to medical suspicion that they have the sickle cell disease, showing that in most cases of the disease are only diagnosed after the appearance of the first signs and symptoms (Figure 4).

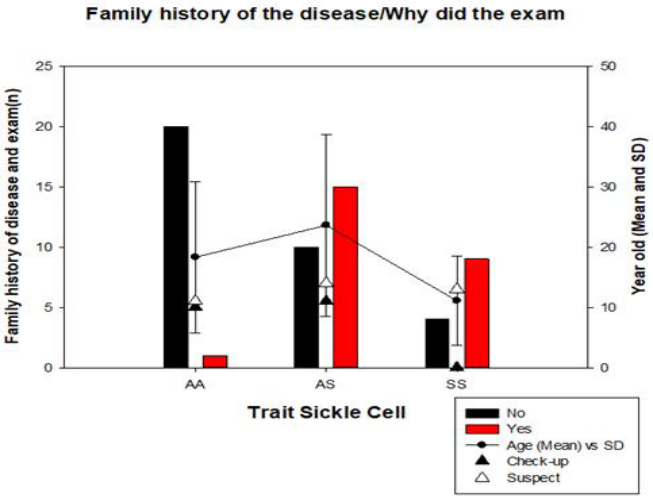

Figure 4: Family history of disease and reason for electrophoresis. **(AA)**: group of participants who, when submitted to hemoglobin electrophoresis, the results did not present the sickle cell trait in any of the alleles (Hb AA). **(AS)**: groups of participants who, when submitted to hemoglobin electrophoresis exam, the results of the exams presented an allele with the sickle cell trait (Hb AS). **(SS)**: group of participants who, when submitted to electrophoresis examination, presented the sickle cell trait in both alleles (Hb SS). Related to the history of the disease in the family and whether the indication for the hemoglobin electrophoresis test was due to suspected sickle cell anemia or just routine tests.

It was found that there is little knowledge about sickle cell anemia and sickle cell trait in our Angolan population, since of the 59 study participants, 41/59 (69.5%) do not know what sickle cell anemia is, especially participants without sickle cell trait (Hb AA) and with heterozygous sickle cell trait (Hb AS), yet about 62% (8/13) of participants with homozygous sickle cell trait or their families did not know what the disease was, demonstrating that in addition to the lack of information that is

common In the Angolan population, the signs and symptoms of the disease are the factors that lead patients to the diagnosis, since at birth in Angola there are no tests such as the foot test that could diagnose sickle cell anemia or sickle cell trait. However, the participants who gave some distinction to the disease were mostly people with sickle cell trait or sickle cell disease, certainly because of their living with the disease or with some sickle cell disease patient in the family (Figure 5).

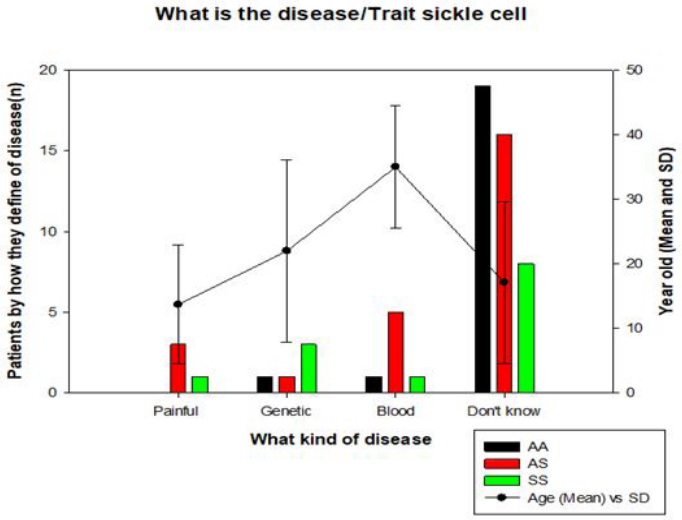

Figure 5: Family history of disease and reason for electrophoresis. **(AA)**: group of participants who, when submitted to hemoglobin electrophoresis, the results did not present the sickle cell trait in any of the alleles (Hb AA). **(AS)**: groups of participants who, when submitted to hemoglobin electrophoresis exam, the results of the exams presented an allele with the sickle cell trait (Hb AS). **(SS)**: group of participants who, when submitted to electrophoresis examination, presented the sickle cell trait in both alleles (Hb SS). Related to the history of the disease in the family and whether the indication for the hemoglobin electrophoresis test was due to suspected sickle cell anemia or just routine tests.

What they know about the disease. **(Painful)**: group of participants who stated that sickle cell anemia is a disease that causes many pains in bones and joints. **(Genetic)**: participants than participants who stated that sickle cell anemia is a genetic disease. **(Blood)**: groups of participants who stated that sickle cell anemia is a disease that causes changes in the components of blood circulation. **(Don't Know)**: group of participants who did not know how to define sickle cell anemia. Related to the number of participants without the sickle cell trait (AA), the sickle cell trait (AS) and sickle cell disease (SS) and the average age for each group.

Discussion

The results of the study showed that the indication for the electrophoresis test for the diagnosis of the sickle cell trait occurs mostly at ages below 20 years, and participants with homozygous for the sickle cell trait (Hb SS) are diagnosed mostly among individuals aged between 0 to 10 years of age, and this condition for some cases extends up to 20 years, which is the period when the first signs and symptoms of the disease begin to occur or problems intensify and requires patients to search for diagnosis, however, the average age of participants with homozygous sickle cell trait was 9 years (SD = 7.4), demonstrating that the diagnosis of sickle cell deseaseoccurs mainly in childhood (Table 1). It was observed that the highest average age among study participants with heterozygous for sickle cell trait (Hb AS) who had an average age close to 24 years old, ie the age that most Angolans begins to form a family, and cross-referencing between individuals with sickle cell trait (Hb AS) is a condition that may favor an increase in the number of sickle cell disease patients, especially in a country like Angola where there is no genetic counseling services for couples who want to have children. There was no very large difference between the number of men and women with sickle cell trait or sickle cell disease.These results differ from a study carried out in Brazil, at the Porto Alegre hospital, where 94 individuals were examined and 47 were patients with sickle cell hemoglobinopathy, and the age was 22 years (± 16.8) [17] and a study in the Santa Casa de Misericórdia of São Paulo in a population consisting of 53 patients, where the median age was 30 years [18], while the average age in our study was close to 19 years (SD = 13.5).The data in the present study is similar to the study carried out in Uganda on 140 children with homozygous SS, where 68% were between 8 and 12 years old and the average age was 14.25 years [19]. These results can confirm the data that pointed out that in 2010, sub-Saharan Africa represented 75.5% of the global number of newborns with sickle cell disease, where the majority of children died before the age of 5 due to a multitude of socioeconomic factors and the lack of assistance in the public health system [19,20].

It was observed in the study a greater number of women than men, 38/21 (64%) and belonged to blood group O with 24/59 (40%), followed by group A with 17/59 (28%) and B with 13/59 (22%) and AB with 6/59 (10%), preliminary studies developed by our study group show that blood group O is predominant among Angolans (Table 1), this result is similar to a study carried out on 10,116 volunteers who donated blood at the national blood transfusion center in Mauritania, in which the overall results showed that group O had the highest frequency (49.10%), followed by A (28.28%), B (18.56%) and AB (4.05%) [20]. The Rh (Rhesus) factor demonstrated that positive Rh (D) people were the majority in relation to negative ones, representing 49/59 (83%) of the studied population (Figure 2) and these data are somewhat similar to those obtained in the study Mauritania, where the Rh+ (positive) factor was by far the most prevalent in 94.23% of the volunteers, while Rh- (negative) was present in only 5.77% of the total population [14,20].

The frequency of the sickle cell trait among the 59 study participants was distributed as follows, 67% (2/3) in the O-Rh(-) group, 67% (2/3) in the B-Rh(-) group, 61% (8/13) in the A-Rh (+) group, 33% (7/21) in the O-Rh(+) group, 33% (2/6) in the AB-Rh(+) group and 30% (3/10) in the B-Rh(+) group.Blood groups when associated with Rh factor and the results of screening for sickle cell trait, the frequency of participants with homozygous sickle cell trait (Hb SS) for the groups were distributed as follows, 50% (3/6) for the AB-Rh(+) group, 33% (1/3) for the A-Rh(-) and B-Rh(-) group, 28% (6/21) for the O-Rh(-) group and 20% (2/10) B-Rh(+) group and there was no presence of the disease (homozygote for sickle cell trait HB SS) in the A-Rh(+) group consisting of 13 participants and in the O-Rh(-) group consisting of 3 participants (Figure 1). These results are different from the study carried out in the collection and transfusion unit of the Hemotherapy Service of Primavera do Leste in Brazil, in 2,708 blood donors and when analyzing sickle cell disease combined with the ABO system and the Rh factor, they found that there was a predominance of individuals class O-Rh+ (0.74%), followed by class A-Rh+ (0.59%) and B-Rh+ (0.48%), while classes AB-Rh+ and AB-Rh- 0.03% and 0.03%, respectively [21].

Most of the homozygous (Hb SS) for the sickle cell trait 62%

(8/13) who participated in the study were adolescents who attended school and reported that due to the signs and symptoms of the disease they have a low school performance and about 31% (4/13) did not perform any type of student or work activity (Figure 2) because they often have to miss school or other daily activities due to the signs and symptoms of the disease. These data had already been reported in the study carried out in Uganda in which the authors stated that there is a scarcity of studies on how the disease impacts on school illness, emotional, physical and social well-being of patients, although its high prevalence in the population and in their study all these items were found to have very low rates in adolescents homozygous for the sickle cell trait who participated in the study and the children's caregivers stated the same when asked about the subject [13].

When asked about the problems that most afflicted them and made them go to the hospital frequently, even without a confirmatory diagnosis of the disease, joint pain was mentioned by all homozygous(HB SS) participants for the sickle cell trait (13/13), by 32% (8/24) of heterozygous (Hb AS) participants for the sickle cell trait and only 5% (1/21) of the participants without the sickle cell trait (Figure 3), this result is similar to the study relaxed in Uganda, where the majority of children 91/140 (65 %) came to the hospital with fever and 89/140 (64%) with pain[13].Anemia was mentioned in about 69% (9/13) of the participants with sickle cell trait, 52% (13/25) of the participants heterozygous(Hb AS) for the sickle cell trait and 57% (12/21) of the participants without the sickle cell trait (Figure 3), these data differ slightly from the study carried out in Sudan, where individuals with sickle cell disease (Hb SS) had a low mean hemoglobin 7.0 ± 1.1 g/dL compared to individuals with sickle cell trait (Hb AS) that presented hematological parameters similar to normal individuals (Hb AA) [3].

It was found that although some of the participants without the sickle cell trait (HbAA) and heterozygous for the sickle cell trait (HbAS) have cited symptoms similar to those mentioned by the homozygous participants for the sickle cell trait (HbSS), while in the homozygous sickle cell trait participants in addition to the drugs cited they also received treatment with hydroxyurea (4/13), persantin (4/13) and transfusion (2/13), even without having a confirmatory diagnosis of the disease (Figure 3). This result is similar to that observed in the Uganda study, where only 35% of participants were treated with hydroxyurea, 131 (95%) were treated with folic acid and 3% received blood transfusions in the last year [13]. A randomized study in children with sickle cell anemia, showed that blood transfusions, analgesia and antibiotics are typified to alleviate specific disease symptoms, while the preventive approach uses pneumonia and influenza vaccination, hydroxyurea to induce fetal hemoglobin (HbF) and transfusions of blood to prevent episodes of primary and secondary stroke [22], we believe that among other factors the deficient economic condition in which the majority of the population lives in Angola is a determining factor so that not all patients take the most appropriate medication for their health condition.

When questioning the existence of people with sickle cell anemia or sickle cell trait in the study participants' family, it was observed that 69% (9/13) of the homozygote (Hb SS) for the sickle cell trait, 60% (15/25) of the heterozygote (Hb AS) for the sickle cell trait and only 5 % (1/21) knew about the existence of a close family member with the disease and this data showed that people with family members with sickle cell anemia should be more attentive and seek the diagnosis earlier to minimize the risks of being diagnosed in a more advanced stage where treatment may be inefficient.This information was sufficient to understand why all (13/13) of homozygote sickle cell trait participant and only 14/25 (56%) of the heterozygousparticipants for the sickle cell trait and 11/21 (52%) of the participants without the sickle cell trait responded that they had the electrophoresis test for suspected sickle cell anemia (Figure 4). We agree with study that claim that in Sub-Saharan Africa, it is estimated that more than 250,000 babies with SS disease are born each year, identifying the genotype with education and counseling about the disease would allow the choice of partner and avoid the risks of the disease and a voluntary pre-marital screening program cut the number of babies born with the disease in half and therefore screening became mandatory [23].

It was found that about 73% (43/59) of the participants did not know what sickle cell anemia is, even among those who suffer or live as a homozygote sickle cell trait participant, about 62% (8/13) did not know how to distinguish the disease and this demonstrate that the country's health services have not provided adequate information to the population about the disease and this contributes to the proliferation of the disease and, in a way, to the worsening of the patient's health condition, considering that the majority of the population in the country is only carrying the sickle cell trait as shown in the present study, it is possible that the number of cases of the disease is still underestimated (Figure 5). This condition helps to maintain the idea that the greatest burden of sickle cell anemia is seen in sub-Saharan Africa, where more than 75% of all sickle cell diseases occur and this proportion is expected to increase by 2050, with a prevalence that varies markedly between different regions, but reaches levels of up to 40% in some areas of sub-Saharan Africa, eastern Saudi Arabia and central India [13,14,15].

Limitation of the study

The study had the limitation to present a number of only 59 participants and this happened because although the study was developed in a single laboratory in a period of one of six months, the electrophoresis test is relatively expensive and many of the individuals with suspected disease are not is able to pay for the exam and, therefore, the results found in this study cannot be representative for the population of Luanda or the country. The other limitation is that with this number of participants there was no way to use inferential statistics, so we had to use only discretionary statistics.

Conclusion

The incidence of sickle cell trait was high among study participants and although the number of participants is small to estimate the incidence of sickle cell anemia in the Angolan population, these results give us the perception that the incidence of the disease and trait is high among individuals suspects, especially among those with a family history of the disease, the blood group O-Rah (+) and AB-Rah (+) appeared to be the most prevalent among participants with sickle cell anemia, frequent joint pain seems to be a characteristic sign of sickle cell anemia and can be used to assess the need to order the hemoglobin electrophoresis test. Further studies are needed to assess not only the incidence of the disease, but also the quality of life, the efficiency of treatment, survival and many other aspects related to sickle cell disease in the Angolan population and that this information be passed on to the population to reduce the incidence of disease in the population.

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