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Original Research Article

Epidemiological and Clinical Profile of Sickle Cell Patients in the Bla Health District

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Abstract

To study the epidemiological-clinical profile of sickle cell patients in the health district of Bla. ***Method:*** This was a prospective and descriptive study over 1 year, from February 2021 to January 2022; relating to all patients who were screened with the TDR_Sikle Scan and/or followed up during the study period. ***Results:*** We performed the TDR in 982 patients from all the consultations (N= 2976). The mean age was 29.6 ± 3.8 years. SS and SC sickle cell disease represent 8.04% (79 cases). Children 23.52% (28 patients) and women 64.35% (51 patients). SS married couples represented (13.92%). Anemia was the most common reason for consultation (47%), followed by bone and joint pain (29%). 33% of the patients (26 cases) had a complication, of which 15 patients had had a CVO and 6 patients (23%) a worsening of the anemia. We have recorded 1 death. ***Conclusion:*** regular medical follow-up of sickle cell patients considerably improves the vital prognosis. The frequency of sickle cell traits remains high including SS and SC forms.

Keywords: Sickle, cell, disease, Hémoglobine, CVO, Anemia, Reference, Heath, center, Bla.

INTRODUCTION

Sickle cell disease is an autosomal recessive hereditary genetic disease. Genetic disease most prevalent in the world. About 7% of the world's population carries an abnormal globin gene [1, 2]. It is a qualitative anomaly of hemoglobin which consists of the replacement of one amino acid molecule by another. For the S and C genes, glutamic acid, in position 6 of the beta chain of globin, is replaced by a molecule of valine and lysine.

It is a hereditary condition that usually manifests after the age of 6 months, transmitted in simple autosomal recessive mode by hemolytic anemia and by an attack of abdominal, back and extremity pain. The diagnosis of hemoglobinosis is possible by highlighting the S, C and F genes contained in the red blood cells. For this diagnosis, hemoglobin electrophoresis and/or other hemoglobin identification techniques including rapid diagnostic test, Sickle Scan, and hemotype are used.

In Africa, it constitutes a real major public health problem, with a proportion of carriers of the S gene between 25 and 40% and that of the C gene between 8-15% [3, 4]. According to the geographical distribution map of the sickle cell gene, the maximum concentration is located in West Africa and Central Africa. Recent global estimates predict over 400,000 major sickle cell syndrome (MDS) births/year in 2050. [5, 6]

In the United States, in the African-American population, one in 600 children is born with the severe form of the disease. [5]. In Gabon, carriers of the sickle cell gene represent 25% of the population. [7]

Several studies have contributed to evoking the role of endogamy and the outbreaks characteristic of certain ethnic groups. [8]

In Mali, statistics show a prevalence of the sickle cell gene at rates of between 6 and 16% of the population in its heterozygous form depending on the region and an annual number of sickle cell births of between 5,000 and 6,000. [5]

The present study is motivated on the one hand by the need to establish the local prevalence of S and C genes in rural areas after patient screening in the health district of Bla and on the other hand by the absence of a of data and a codified support system.

METHOD

This was a prospective and descriptive study over a year from February 2022 – January 2022. Included were all patients screened voluntarily or cases suspected by the TDR sickle Scan in the health district of Bla.

Data analysis was performed using SPSS software.

RESULTS

Sickle cell patients represented 8.04% (N=79) of all people screened (N=982), and 2.65% of all consultations (N=2976). Screening was voluntary in 314 patients (31.97%) of all people screened. The female gender predominated with a ratio of 1.1. Children represented 23.52%, 64.35% of women and 12.11% of men. The majority ethnic groups were essentially the Djonkas (36.04%), the Bobo (23.93%), Minianka (18.02%).

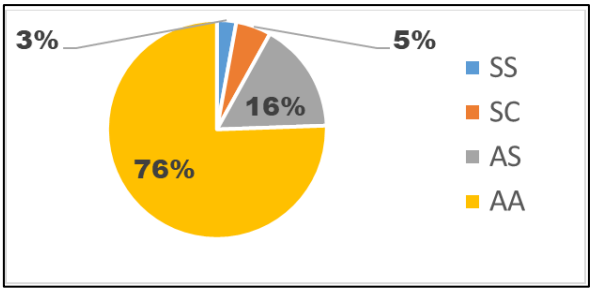

Fig.1 : Frequency of profiles

Table I: Age

Age	Frequency	Percentage
1 – 5 years	14	17,72%
6 – 12 years	6	7,59%
13 – 20 years old	9	11,39%
21 – 35 years old	27	34,17%
>35 years	23	29,11%
Total	**79**	**100%**

The average age was 29.6 ± 3.8 years with extremes of 4 and 47 years. The 21-35 year old age group was the most represented, ie 34.17% of patients, of whom 8 patients had SS form. In our study, The marital status of sickle cell patients revealed that 11 patients (13.92%) patients with SS form were married, 21 patients (26.58%) with SC form cohabiting and 47 patients (59.49%) were single, 17 of whom had SS form. Nineteen patients (24.05%) were from a consanguineous marriage and 26 patients (32.91%) had a partner with sickle cell trait. Four patients (5.06%) had a higher education level, 49 (62.02%) had a professional activity including 7/49 (14.28%) in unstable activity.

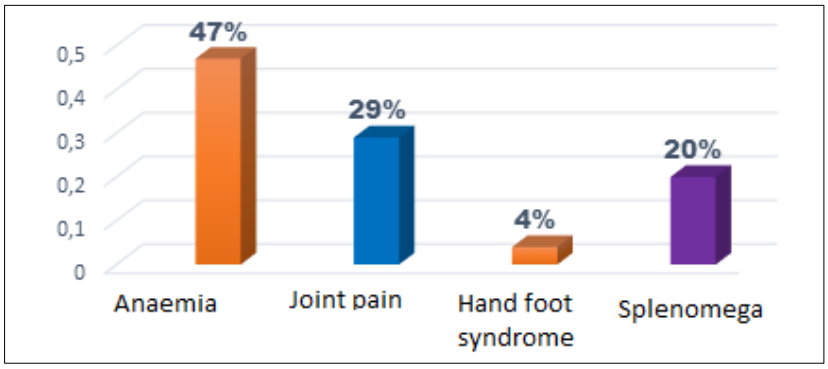

Fig. 2: Reasons for Consultation

Anemia was the most common reason for consultation, at 47%, followed by osteo-articular pain (29%).

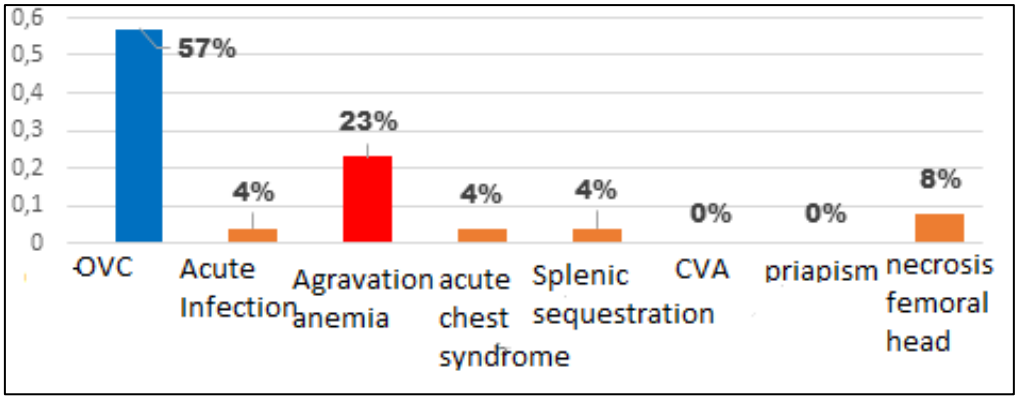

Fig.3 : The Complications

Hand-foot syndrome (sickle cell dactylitis) was observed in 14 patients (17.72%) whose age was between 3 and 5 years old. Splenomegaly accompanied anemia in 3 patients including 1 AS form and 2 SC cases. Asplenia was suspected in an SS patient who was frequently admitted to the service in an array of recurrent infectious syndrome. No cardiomyopathy and/or cholelithiasis were found.

During the study period, 33.91% of patients (26 cases) presented at least one complication and 53 patients (67.08%) presented no major complication. Fifteen patients (57%) had a CVO, and 6 patients (23%) an aggravation of anemia including 1 pregnant woman with SC form, 1 case of ATS and 2 cases of splenic sequestration all with SC form. Vaso occlusive crisis, generally at the forefront of the disease, 4 patients with SS forms had had a similar attack twice in the month and 11 patients, 8 of whom had SC form, were in their first episode.

We observed 5 patients in consultation for lameness, including 2 cases (1 SS case and 2 SC cases) of confirmed osteonecrosis of the femoral head and 1 case associated with painless splenomegaly. Sixty-six patients (83.54%) had a hemoglobin level between 6 – 9g/dl with an average of 7.2g/dl. We performed a transfusion of red blood cell concentrate in 29 patients (36.70%) and the transfusion exchange was performed in 2 patients. We have recorded 1 death.

All patients had received maintenance treatment with folic acid for 10 days per month, compressed zinc for 5 days per month and sulfadoxine perymethamine in a single dose per month depending on their weight and age. Hydroxy urea was not used in any of our patients. In our study, 77.21% or 66 patients had regularly undergone maintenance treatment (medical follow-up) and 13 patients (16.45%) had not been able to carry out all the assessments necessary for their follow-up.

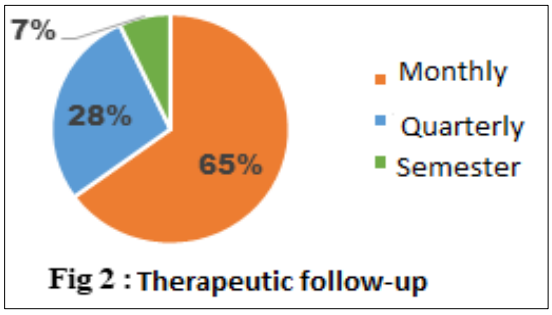

Fig 2 : Therapeutic follow-up

DISCUSSION

The prevalence of the sickle cell gene in the Ségou region is 13.8%. [5] Our frequency was statistically lower than that of THIERO TA et al. P=0.008. This difference could be explained by the size of the study population which is greater than that of our study.

Our majority age group of 21-35 years is comparable to that of the Marielle and Aissata study without statistical difference [1, 9] P>0.05. The juvenile period seems to be the period of discovery of the disease in our context, because the multiple symptoms of the disease are generally confused with other infantile and/or tropical pathologies. Female predominance has also been observed in other series [10, 11, 12]; on the other hand, the male sex was frequently observed in the study by Aissata, [9].

Since sickle cell disease is not a sex-linked autosomal disease, the observations in the various studies could only be explained by the method of recruitment and the high female density in the population. The normal AA phenotype, frequently observed followed by the sickle cell trait AS in our

study, was statistically comparable to that of Thiéro [9], most of which was screened voluntarily.

The hand-foot syndrome (sickle cell dactylitis) occurs in 10 to 20% of children with sickle cell disease under four years of age [13], which is consistent with our study (17.72%) P>0.05. The frequency of moderate anemia was without statistically significant difference with that of the Aissata study which found 70.8% of cases of anemia. P=0.78. The low average hemoglobin level in our patients was discovered during the consultation which discovered their profile for which no follow-up was initiated. This was also observed in other series where the average rate was between 5.6 - 9.5g/dl. [9, 11]

No case of cholelithiasis was observed in our study, unlike Diop who in his study reported a predominance of cholelithiasis in 10% of cases [14]. Ischemic complications, in particular aseptic osteonecrosis of the femoral head in our case, were statistically lower than that of Aissata, who found 17% cumulatively (bone and ocular) [8] P = 0.037; Cabannes [15] reported a predominance of these two types of ischemic complications.

The main acute complications (vaso-occlusive crisis, acute chest syndrome, sudden anemia, severe infection, stroke) can occur unpredictably in patients with sickle cell disease and jeopardize their vital prognosis. This does not require a diagnosis and an urgent therapeutic management. No case of vascular accident was observed. The occurrence of painful crises in our patients would be linked on the one hand to ignorance of their status and on the other hand by the lack of medical follow-up which could reduce its occurrence. [8, 9].

Twenty-nine patients (36.70%) were transfused to correct the severe anemia on the one hand and to compensate for the blood loss caused by bleeding on the other. The management of CVO was essentially based on sufficient hydration and analgesics; transfusion was proposed in the event of a prolonged and persistent attack and/or severe acute anemia. The pregnant woman had discovered her status at 27 SA. The prenatal consultations had made it possible to carry out a prophylactic cesarean delivery at 38 weeks. The postoperative follow-up was simple.

A monthly follow-up program was offered to all patients, among whom 31 patients had been able to follow the follow-up regularly for 6 months. The follow-up of the patients in our context was essentially based on the observance of the treatment and the respect of the hygiene-dietetic advices. The death occurred in a patient in a context of worsening anemia associated with osteonecrosis of the femoral head.

CONCLUSION

Early management and regular medical follow-up considerably improve the vital prognosis. The frequency of sickle cell traits remains high including the SS and SC forms. Communication on the transmission mechanisms of the disease could considerably reduce its prevalence.

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