SAS Journal of Medicine
Abbreviated Key Title: SAS J Med
ISSN 2454-5112
Journal homepage: https://saspublishers.com

OPEN ACCESS

Pediatrics

Epidemioclinical Aspects of Sickle Cell Disease in the Pediatrics Department of Sikasso Hospital

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DOI: 10.36347/sasjm.2024.v10i04.006 | **Received:** 28.02.2024 | **Accepted:** 06.04.2024 | **Published:** 16.04.2024

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Abstract **Original Research Article**

Sickle cell disease still constitutes a worrying public health problem today. In its major form, it remains a significant cause of infant mortality in our countries. ***Objective***: To study the epidemiological aspects of sickle cell disease in the pediatric department of Sikasso hospital. ***Methodology***: This is a prospective cross-sectional study running from January 1, 2017 to December 31, 2017, a period of one year, meeting the inclusion criteria. ***Results***: During the study period, 4800 patients were consulted in the department including 72 cases of sickle cell anemia, i.e. a prevalence of 1.5%. The average age was 29.5 months. Children under 5 years old were the most represented with 55.5% of cases. The male sex was more common with a sex ratio of 1.4. Depending on the father's Hb type, the AS form represented 87% of cases. In 14 cases, or 70%, the mothers of our patients carried the sickle cell AS trait. Patients were known to have sickle cell disease before the age of 5 in 55.5% of cases. The SS form was the most represented with 57% of cases. The circumstances of discovery were dominated by abdominal pain and osteoarticular pain with 41.7% and 27.8% respectively. Pallor and jaundice were the most common signs on admission with 37.5% and 17.5% of cases respectively. Complications were dominated by CVO and infection with respectively 56.2% and 25%. ***Conclusion***: Difficulties remain in management, linked on the one hand to the unfavorable socio-economic conditions of our populations and on the other hand to the unavailability of certain key tests such as hemoglobin electrophoresis and reticulocyte levels at Sikasso hospital.

Keywords: Sickle cell disease, child, clinic, paediatrics.

INTRODUCTION

Sickle cell anemia is an autosomal recessive genetic disease characterized by mutation of a hemoglobin beta chain gene. This mutation induces the synthesis of an abnormal hemoglobin called hemoglobin S (HbS) [1].

It is an autosomal codominant hereditary disease [2]. The hemoglobin that results from this sequence of amino acids is called sickle cell hemoglobin or hemoglobin S (HbS). The inheritance of sickle cell disease is autosomal and recessive. In Africa, it is particularly prevalent in Sub-Saharan Africa where the prevalence of sickle cell trait in the general population sometimes exceeds 30% [3].

In Mali, approximately 12% of the population carries the sickle cell trait and 5000 to 6000 children are born with a major sickle cell phenotype per year [4]. The period from 6 months to 5 years characterized mainly by serious infectious complications responsible for frequent hospitalizations and significant mortality, often fatal splenic sequestration accidents.

Since the creation of the pediatric department at Sikasso hospital, no study has focused on the epidemiological aspects of these children with sickle cell

disease. This motivated us to carry out this work by setting ourselves the following objectives: Study the epidemiological aspects of sickle cell disease in the pediatric department of Sikasso hospital, to try to answer the following questions.

PATIENTS AND METHOD

We conducted a study at the pediatric department of Sikasso hospital. Sikasso region is the third administrative region of Mali, it occupies the south of the national territory. It has an area of 76,480 km2 or 5.7% of the national territory and has a density of 34 inhabitants/km2. The population is estimated at 2,625,919 inhabitants, or 18.1% of the Malian population (INSTAT, 4th RGPH April 2009). This was a prospective cross-sectional study ranging from January 1, 2017 to December 31, 2017, a period of one year. The study focused on all children aged 0 to 15, known to have sickle cell disease, followed in the department during the study period. All children whose sickle cell disease was not confirmed by Hb electrophoresis were excluded from the study. The data collected was using a pre-established survey form. Data analysis was carried out using IBM SPSS 20 software while respecting the principles of confidentiality. Informed consent was obtained from the parents of all patients.

RESULTS

Sociodemographic aspects:

Frequency: During study period, 4,800 patients were consulted in the department, including 72 cases of sickle cell disease, i.e. a prevalence of 1.5%. Children under 5 years old were the most represented with 55.5% of cases. The average age was 29.5 months. The male sex (43/72 ¬) was the most common with a sex ratio of 1.4. The fathers of our patients were not educated in 52.7% of cases. Depending on the Hb type of the father, the AS form represented 87% of cases, and in 70%, the mothers of our patients carried the AS sickle cell trait. The consanguinity relationship in marriage was found in 34.7% of cases. We found that in 22.2% (N= 16) children have at least one sibling with sickle cell disease.

Clinical aspects:

Clinically Patients were known to have sickle cell disease before the age of 5 in 55.5% of cases. In terms of circumstances of discovery, abdominal pain and osteoarticular pain were dominated with 41.7% and 27.8% respectively; and hand-foot syndrome 8.3% (N= 6); detailed in Table I: discovery circumstance. The SS form was the most represented with 57% of cases. Pallor and jaundice were the most common signs with 37.5% and 17.5% of cases respectively. Patients did not benefit from blood transfusion in 86.2% of cases. Patients were correctly vaccinated according to the expanded vaccination program in 91.7% of cases.

Table I: Distribution of patients according to age

Age in months	Frequency	Percentage
0-59	**40**	**55,5**
60-119	21	29,2
120-180	11	15,3
Total	72	100

Children under 5 years old were the most represented with 55.5% of cases. The average age was 29.5 months.

Table II: Distribution of patients according to consanguinity in marriage

Inbreeding	Fréquency	Percentage
Yes	25	34,7
No	**47**	**65,3**
Total	72	100

The consanguinity relationship in marriage was found in 34.7% of cases.

Biological aspects:

Hemoglobin electrophoresis revealed (41/72) cases of SS form that represent 57%; SC 33.3%, Sβ 9.7%. Patients had a hemoglobin level greater than or equal to 6g/dl in 86.2% of cases during attacks.

Evolution and future of patients:

In our study, outpatient treatment was carried out in our patients in 72.8% of cases and for hospitalized patients the duration of hospitalization was less than or equal to 3 days in 66.8% of cases. Complications were dominated by vaso-occlusive crises (VOC) and infection with 56.2% and 25% respectively. We recorded only one case of death due to acute chest syndrome (ATS).

DISCUSSION

Sociodemographic aspects:

From January 1, 2017 to December 31, 2017, we collected 72 files of children with sickle cell disease out of the 4,800 patients consulted in the pediatric department of Sikasso hospital, i.e. a frequency of 1.5%. This frequency is lower than that of DIONE L [5] which obtained a frequency of 27%. This difference is due to the fact that the CHU Gabriel Touré is in center of capital therefore more accessible, and due to the density of the population. Children aged 0-59 months (0-4 years) were most represented with 55.5%, and mean age was 29.5 months. This result is similar to that of DOUMBIA A [6] who found 48%. This could be explained by the fact that around 12 to 48 months, HbS almost completely replaces fetal hemoglobin (HbF), hence the high frequency of crises and even complications (in particular infections and anemia). and that from 8-9 years old, it begins to decline. The result of Diarra Y *et al*., is superior to our result with an average age of 84 months [7]. The male gender was more represented (59.2%) than the female gender with a ratio equal to 1.4. DOUMBIA A [6] and FOFANA D [8] found predominantly male results with a sex ratio of 1.54 and 1.28 respectively. Mali remains a

country rooted in its traditions and customs, and endogamy is proof of this. Many authors have cited the latter as a risk factor for the disease [9, 5, and 10]. We found almost 34.7% of the children in our study were born from a consanguineous marriage. We found that 22.2% of cases have at least one sibling with sickle cell disease. DOUMBIA A [6] found 43.3% of cases with at least one sickle cell brother and ELOUNDOU CO [11] found 35% of cases with at least one sickle cell brother.

Clinical aspects:

The circumstances of discovery:

Abdominal pain was the most cited discovery circumstance with 41.7% of cases followed by osteoarticular pain 27.8%. This result is different from that of DOUMBIA A [6] who found osteoarticular pain, hand-foot syndrome and infections as circumstances of discovery with respectively 30%, 25.3%, and 19.3% of cases. A third of the patients in our series presented hand-foot syndrome as their first attack before reaching their fifth birthday. These results are less important than those of SHONGO who found in the series 56.1% of hand-foot syndrome as the first sickle cell crisis and 82.9% of children having presented their first crisis before reaching their first birthday [6]. Pallor and jaundice were the most common signs with 37.5% and 17.5% respectively. This could be explained by the hemolytic nature of sickle cell disease. Our results are close to those of DOUMBIA A [6] who found 35.3% and 16.6% respectively for pallor and jaundice. FOFANA D [5] found a predominance of jaundice and NEHOULNE G [12] a predominance of fever.

Biological aspects:

In our study, the blood count was systematically performed for patients. Anemia was found in all children. The baseline hemoglobin level was on average 8.8g/dl in the homozygotes, similar to that of I Diagne *et al*., [13] where the baseline hemoglobin level is 8.27 ± 1.36 g/dl in the homozygous. A study carried out in Burkina Faso found a median total hemoglobin (Hb) level at inclusion was significantly higher for HbSC children compared to HbSS (9.5 g/l versus 7.0 g/l) [14]. The anemia was normochromic, normocytic in more than three quarters of cases (76.4%) and regenerative in 69.4% of cases. It is explained by chronic hemolysis in children with sickle cell disease. More than half of our sickle cell patients 52.7% (N= 38) had a hyperleukocytosis greater than 10,000 leukocytes/mm3. with an average of 12,465 elements/mm3. Indeed, hyperleukocytosis is physiological in sickle cell disease and could be explained by the cause's hyperactivity of the marrow and inflammatory phenomena [15, 16]. Similar results are found in the literature [17].

Evolution and future of patients:

We recorded 16 cases of acute complications among patients, or 22.2%. In 56.2% (n = 9) of cases, presented a CVO. This result is lower than that of DIALLO D [8] and FOFANA D [7] who found 69.7% and 65.2% CVO respectively. DOUMBIA A [6] found 36.7% in favor of infections and 30% in favor of CVO. We deplored the death of a 6-year-old SS patient due to acute chest syndrome (ATS).

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