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Epidemiological, diagnostic and evolutionary aspects of sickle cell disease in children at the University Hospital of Libreville

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Abstract

Introduction: The aim of this work was to determine the epidemiological and clinical aspects of children with sickle cell disease hospitalized in the pediatric department of the University Hospital of Libreville (CHUL).

Method: This was a retrospective, descriptive and analytical study of the files of children with sickle cell SS hospitalized in the pediatric department of CHUL, a reference hospital in Libreville and Gabon, from October 2022 to September 2023.

Results: the frequency of sickle cell patients hospitalized was 9.9% [8.8% - 11.1%]. Of the 201 files selected, 125 (62.2%) were boys, i.e. a sex ratio of 1.64. The average age of the children included was 72.3 ± 52.7 months, the minimum was 6 months and the maximum 208 months. The mothers of the hospitalized children were without gainful activity in 62.4%. The mean hemoglobin level on admission was 5.7 ± 2 g/dl. Vaso -occlusive crisis was found in 51.7%, malaria in 18.9% of cases. The mean length of hospitalization was 6.1 ± 3.9 days, bronchopneumopathies were the pathologies with the longest hospitalization durations ($p<0.01$). The evolution was marked by death in 1.5% of cases, these deaths were associated with a median hemoglobin level on admission of 1.9g/dl.

Conclusion: sickle cell disease is a common cause of hospitalization in our context. The mortality rate of sickle cell patients requires an exploration of the epidemiological factors impacting the criteria on admission.

Keywords: Sickle Cell Disease; Children; Associated Factors; Hospitalization; Libreville

1. Introduction

Sickle cell disease is an autosomal recessive genetic disorder characterized by the mutation of the gene coding for the beta chain of hemoglobin. This mutation induces the synthesis of an abnormal hemoglobin, called hemoglobin S (HbS) [1]. Sickle cell disease is the most widespread genetic disorder in the world, according to the World Health Organization (WHO) nearly 5% of the world population carries the sickle cell trait. Some populations in sub-Saharan Africa see this prevalence exceed 30% [2]. In Gabon, the frequency of sickle cell trait is 25%, while 1.8% of the population is homozygous, with nearly 800 births of sickle cell children per year [3].

The life of a child with sickle cell disease is marked by the clinical manifestations of the pathology. These clinical manifestations present a great variability in their expression, but also in their severity, leading to a frequent need for hospital care [2].

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In Mali, according to Cissouma et al, children with sickle cell disease represented a frequency of 1.5% of children seen in consultation [4]. Akolly et al found a frequency of 3.22% of sickle cell disease among children hospitalized in Lomé, Togo [5].

In Gabon, despite the prevalence of sickle cell disease in the population, there is little data on the epidemiological and clinical aspects of hospitalized children with sickle cell disease.

The objective of this work was to determine the epidemiological and clinical aspects of children with sickle cell disease hospitalized in the pediatric department of the University Hospital of Libreville (CHUL).

2. Method

This was a retrospective, descriptive and analytical study, covering the files of children with sickle cell SS hospitalized in the pediatric department of CHUL, a reference hospital in Libreville and Gabon, from October 2022 to September 2023.

The study population consisted of all infants, children and adolescents with homozygous SS sickle cell disease. We included children with SS sickle cell disease documented by hemoglobin electrophoresis or HPLC, hospitalized in the department during the study period, regardless of the outcome of the hospitalization. We did not include children with heterozygous AS, SC or homozygous CC, as the files had missing data.

The data collected were: sex, age, date of entry and exit, hemoglobin level on admission, diagnosis, evolution, age and socio-professional category of the mother, affiliation to the National Health Insurance and Social Security Fund (CNAMGS).

We defined as "managers" mothers who are executives in the civil service or salaried executives in the private sector; "masters" mothers who are not executives in the private sector; "executive" mothers in the executive category in the civil service or the private sector or entrepreneurs in the informal sector; "without" mothers with no source of income. Patients registered with the CNAMGS as having a long-term condition (ALD) benefit from a co-payment of 10% of the patient's costs for care related to their ALD, compared to 20% for other care or other patients.

Data were collected on Epi Info 7.2.2©. We performed a descriptive analysis to determine the characteristics of the sample. The age of the children, the hemoglobin level on admission and the length of hospitalization were expressed as mean and standard deviation or median, with calculation of the 1st (Q1) and 3rd (Q3) interquartiles. The medians were compared using the Mann-Whitney or Kruskall -Wallis test when the number of groups was greater than 3. Qualitative data were expressed as frequency, with a 95% confidence interval according to the Miettinen method. To compare frequencies we used the Chi-square test of independence or Fisher when the numbers were insufficient. The statistical significance threshold was set at $p < 0.05$ for a bilateral test. The analysis, tables and figures were performed on MS Excel.

The study received the favorable opinion of the Scientific Committee of the CHUL, and the collection was carried out in strict compliance with the principles of Good Clinical Practice and the Helsinki Declaration 2013 revised in 2018.

3. Results

Figure 1 shows the flow diagram of the cases included in the study. Of the 247 sickle cell subjects hospitalized during the study period, the sex distribution was 143 (57.9%) boys. Of the 201 cases retained, 125 (62.2%) were boys, for a sex ratio of 1.64.

The mean age of the children included was 72.3 ± 52.7 months, the minimum was 6 months and the maximum 208 months.

The mothers of the hospitalized children had a mean age of 32.7 ± 7.6 years, the minimum was 18 years and the maximum was 56 years. The mothers were without gainful employment in 62.4% of cases. The distribution of mothers according to their gainful employment is shown in Figure 2.

Hospitalized children benefited from CNAMGS insurance for 88.1% (n=177), including 8.5% (n=17) registered ALD, and 11.9% (n=24) did not have CNAMGS insurance.

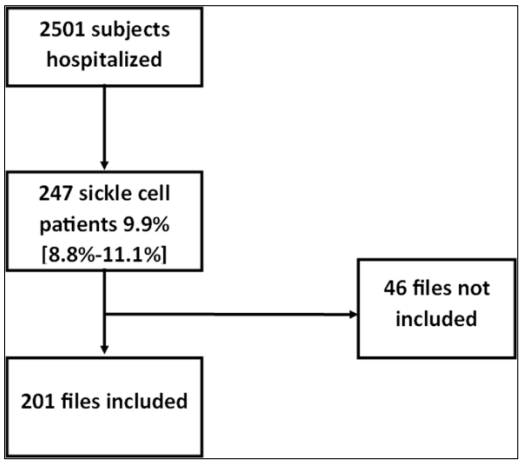

Figure 1 Flow chart of subjects included in the study

The mean hemoglobin level on admission was 5.7 ± 2 g/dl, the minimum 0.9 g/dl, the maximum 10 g/dl. The diagnosed pathologies are classified in Figure 3. The mean duration of hospitalization was 6.1 ± 3.9 days, the minimum 1 day and the maximum 38 days. The patients were discharged from the department to their homes in 98.5% of cases (n = 198), 3 patients died (1.5%).

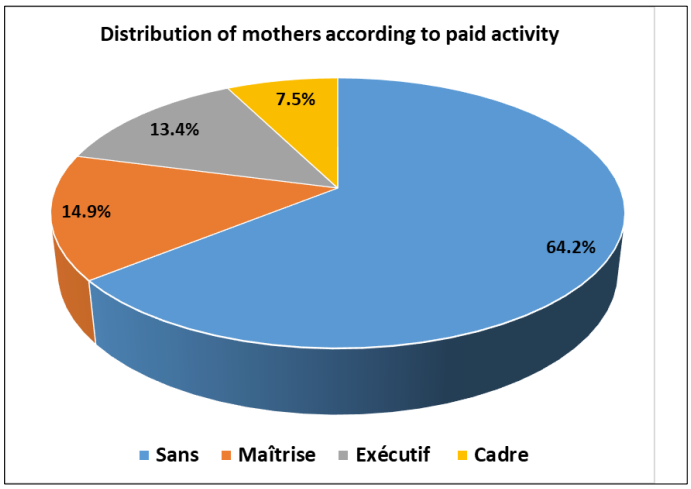

Figure 2 Distribution of mothers according to their paid activity

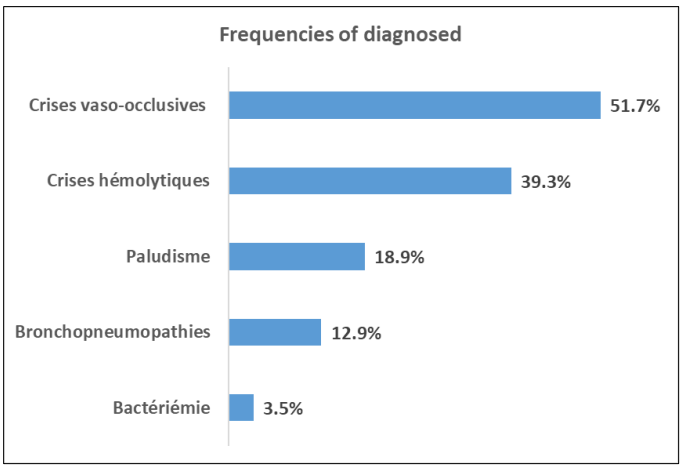

Figure 3 Frequencies of conditions diagnosed in hospitalized sickle cell patients

The analysis between the duration of hospitalization and the pathologies found showed that bronchopneumopathies were the pathologies with a longest duration of hospitalization (table 1). The univariate analysis of the factors associated with the evolution showed that deaths were linked to a median hemoglobin level on admission at 1.9 g/dl (table 2).

Table 1 duration of hospitalizations according to pathologies

Diagnosis	**Median in days**	**[Q1-Q3] days**	***p***
Vaso -occlusive crises	5.5	4 - 7	
Hemolytic crises	5	3 - 5.3	
Malaria	5	4 - 6	
Bronchopneumopathies	8	6 - 10	*< 0.01*
Bacteremia	7	4.8 -10	

Table 2 Factors associated with evolution

Associated factors	**Favorable evolution (n=198)**	**Death (n =3)**	***p***
Age (months), median [Q1; Q3]	60 [36 ; 96]	168 [108;186]	0.091
Hemoglobin at admission g/dl, median [Q1; Q3]	5.7 [4.5; 7.0]	1.9 [1.4; 2.4]	**<0.01**
Mother's activity,			
Without	127 (64%)	2 (67%)	0.55
Mastery	30 (15%)	0 (0%)	
Executive	26 (13%)	1 (33%)	
Frame	15 (7.5%)	0 (0%)	
CNAMGS insurance			
Yes	158 (80%)	2 (67%)	0.5
No	23 (12%)	1 (33%)	
ALD	17 (8.6%)	0 (0%)	
Sex			
Male	122 (62%)	3 (100%)	0.29
Female	76 (38%)	0 (0%)	

4. Discussion

Despite the limitations of our study related to the retrospective nature of the recruitment of files, this study is representative of children with sickle cell disease hospitalized in Libreville. Indeed, the CHUL is the main hospital in Libreville and Gabon. The hospitalized patients come from all categories and social classes, including the working classes [6]. The average age of the mothers was 32.7 years, and 64.2% of them had no gainful activity. This result is up from that of a previous study by Minto'o et al who found a rate of 52.2% of mothers with an income-generating activity and therefore able to contribute directly to household expenses [7]. The difference could be linked to the covid-19 crisis that occurred between these two studies, and which led to a loss of activity especially in the informal sector [8].

Registration with the CNAMGS was effective for 88.1% of hospitalized patients. Only 8.4% had ALD coverage. The proportion of 11.9% of unregistered patients could be patients of non-national parents; compliance with ethical principles did not allow the inclusion of this data in this study. Furthermore, the low rate of ALD declaration patients is

a barrier to care, because it is mandatory from the diagnosis of the pathology, and constitutes an important step forward in overall medical care for Gabon [6].

All children on admission were anemic, the mean hemoglobin level on admission was 5.7 g/dl, this mean is lower than those found by Thiam et al, as well as Koko [9,10]. The difference could be based on the study populations of the different surveys. Thiam and Koko investigated children in the inter-critical period, while we recruited children hospitalized therefore in the middle of a crisis. The diagnoses found were in order of frequency vaso -occlusive crises, hemolytic crises and infectious pathologies. This observation is the one encountered in several other surveys such as that of Mabiala et al in Brazzaville, as well as Koko et al in Libreville, while in Gueï-Couitchere et al in Abidjan hemolytic crises were more frequent than painful crises [9,11,12]. These similarities between the studies of Brazzaville and that of Libreville, and the dissonance with that of Abidjan can be justified in the haemoglobin S haplotype. Indeed, the area of Congo and Gabon is that where the Bantu haplotype is found, known as the one with a greater expression of painful crises, while the Cocody haplotype found in Abidjan presents fewer painful crises [13].

The most frequently encountered infections diagnoses were malaria and bronchopneumopathy. This order of frequency is the same in the general pediatric population, with malaria constituting 40% of pediatric hospitalizations in Libreville [14]. It should also be noted that pneumococcal vaccination is not included in the national vaccination schedule. Furthermore, the duration of hospitalization for malaria was shorter than that of hospitalizations for bronchopneumopathy. One of the reasons for this shorter duration is that the treatment of malaria is standardized, and is given for a maximum of 3 days, while the treatment of bronchopneumopathy requires identification of the germs by culture and an antibiogram that lasts from 3 to 5 days.

The factor associated with the evolution was the hemoglobin level at admission. Indeed, the deceased patients had a median hemoglobin of 1.9 g/dl, while the general median was 5.7 g/dl. The deceased patients therefore arrived at the hospital with a level well below the level defining severe anemia, i.e. < 5g/dl. The causes of these arrivals in such a late context should be investigated in further studies.

5. Conclusion

If sickle cell disease represents only 1.8% of the general Gabonese population, it represents 10% of the hospitalized pediatric population. The profile of hospitalized sickle cell disease patients is marked by painful crisis as the main clinical expression and malaria as the primary cause of hospitalization. The evolutionary profile is essentially conditioned by the hemoglobin level on admission. The reasons for this low hemoglobin level must be sought in a population survey in order to curb this identified factor.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

Ethical approval was obtained.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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