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Study of the Management of Children with Major Sickle Cell Disease in Pediatric Emergencies at the Gabriel Toure University Hospital

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Abstract

Objective: To study the epidemiological, clinical and therapeutic aspects of sickle cell disease in children admitted to the pediatric emergency department of the Gabriel Toure University Hospital. Materials and methods: This was a prospective study over 12 months in sickle cell children aged 6 months to 15 years. Results: The frequency of sickle cell disease was 4.67% with 71.1% of SS form. The age range of 60 - 120 months predominated with 43.4% and the sex ratio 1.4. Vaso-occlusive crises (VOC) were the most frequent reason for consultation (50.6%) and osteoarticular pain was the main symptom followed by fever and pallor with respectively 67.5%, 60.2%, 39.8%. The pain subsided in less than 72 hours in 39.8% and hyper hydration was performed in 85.1% of patients. Analgesic treatment was given in 90.4%, antibiotic therapy in 51.8% and phenotyped red blood cells transfusion was performed in 36.1%. The average duration of treatment was 10 days. Conclusion: Vaso-occlusive crisis remains the main reason for consultation in sickle cell disease children at Gabriel Toure University Hospital. The management consisted, in addition to the correction of the triggering factors, of hyper hydration and the administration of analgesics and blood products.

Keywords

Sickle Cell Disease, Pediatrics, Emergencies

1. Introduction

The situation of sickle cell disease in Mali is characterized by a prevalence of sickle cell trait that varies between 4% and 15% from north to south and an estimated number of sickle cell births between 5000 and 6000 per year, a high mortality rate among sickle cell patients who have no access to specific care: 50% of deaths before the age of 5 [1]. The major form is at the origin of various acute complications whose management constitutes a medical emergency.

That is why we initiated this work to determine the frequency, the clinical manifestations and the therapeutic approach to acute complications of sickle cell disease.

2. Materials and Methods

Our study was conducted in the pediatric emergency department of the Gabriel Toure University Hospital. This was a prospective and descriptive study over 12 months from June 1, 2017 to May 31, 2018, which involved children with sickle cell disease aged 6 months to 15 years whose diagnosis is established by hemoglobin electrophoresis and complete blood count. Were included all children with sickle cell disease admitted in crisis, during the study period. Sickle cell children coming only for their routine follow-up and sickle cell children older than 15 years were not included. Thus, we included 83 children with sickle cell disease in emergency situations. The parents of the children included in the study were given a consent form and confidentiality of data was respected during the study. This study was approved by the Gabriel Toure University Hospital Medical Committee.

To evaluate the intensity of pain: the following were used [2]

- The face scale: for children under 5 years old
- The visual analog scale (VAS): for children aged 6 - 10 years
- The numerical scale from 0 - 10: from 10 years old

Thus we have considered as:

Scale: 0 = no pain, 1 - 2 = light pain, 3 - 4 = medium pain, 5 - 6 = severe pain, 7 - 8 = very severe pain, 9 - 10 = intolerable pain.

Data collection was carried out by means of a questionnaire form filled in from the patient's hospitalization records and the sickle cell patient follow-up book. SPSS software version 16.0 was used for data entry and analysis.

3. Results

We collected 83 records of children with sickle cell disease out of 1777 patients hospitalized in the Pediatric Emergency Department, *i.e.*, a hospital frequency of 4.67%. The age range of 60 to 120 months represented 43.4% with an average age of 94 months and extremes ranging from 8 to 180 months. The sex ratio was 1.4. The peak of hospitalizations was observed in December. Consanguinity in marriage was noted in a quarter of the parents, more than half of the children had a history of familial sickle cell disease (55.4%). The majority of the mothers (53%)

did not attend school. A history of hand-foot syndrome was found in 37.3% of children. The majority of children (89.2%) had a correct vaccination status according to the Malian EPI. Vaccines not included in the EPI were anti-pneumococcal 23 (26.5%), and anti-typhoid (25.3%). Two-thirds of the children (67.5%) had a nutritional deficit. Nearly half of the children had their first attack, *i.e.* 45.78%. More than half of the children (60.2%) had a fever at entry and the mean temperature was 37.89˚C, with extremes ranging from 35.1˚C to 40.2˚C. Respiratory rhythm abnormalities (polypnea) were observed in 86.7% of children with a mean of 35 cycles/min and extremes ranging from 20 and 88 cycles/min. Heart rhythm abnormalities (tachycardia and bradycardia) were observed in 39.8% of children with a mean of 117 beats/min and extremes ranging from 80 to 172 beats/min. Oxygen desaturation was observed in 29% of cases. Mean saturation was 93.75% with extremes ranging from 57% to 100%. Osteoarticular pain was the main clinical sign with 67.5%, followed by abdominal pain (45.8%) and mucocutaneous pallor (39.8%) (Table 1).

The pain was of the pulsatile type in 24.1% of cases and of the twisting type in 19.3% of cases (Table 2).

Table 1. Distribution of patients according to clinical signs.

Clinical signs	Workforce	Percentage
Osteoarticular pain	**56**	**67.5**
Abdominal pain	38	45.8
Mucocutaneous pallor	33	39.8
Signs of respiratory struggle	26	31.3
Chest pain	25	30.1
Conjunctival Jaundice	25	30.1
Rails	16	19.3
Hepato-Splenomégaly	9	10.8
Skin lesions	7	8.4
Heart murmur	2	2.4

Table 2. Distribution of patients by type of pain.

Type of pain	Workforce	Percentage
No answer	**35**	**42.1**
Pulsatile	20	24.1
Twist	16	19.3
Burns	5	6.0
Total	**83**	**100**

At H0 admission, 41 patients (49.4%) had very severe pain, at H72 admission 33 children (39.8%) had no pain (Figure 1).

At admission 54.2% of children had multiple sites of pain (Table 3).

The pain occurred in the morning in 31 children. More than half (53%) of the children did not know why their seizure occurred, intense physical effort and stress were found in 15.7% and 12% of the children respectively. Pulmonary infections (9.6%), osteoarticular infections (3.6%) and malaria (2.4%) were found as triggering factors.

The homozygous form (SS) was the most represented with 71.1% (Figure 2).

Hyperleukocytosis was found in 74.69% of children. Anemia was normocytic in 45.8% and normochromic in 84.4% of children and regenerative (26.5%). Of the 60.3% of children who had a thick blood drop, more than half were positive (38.6%).

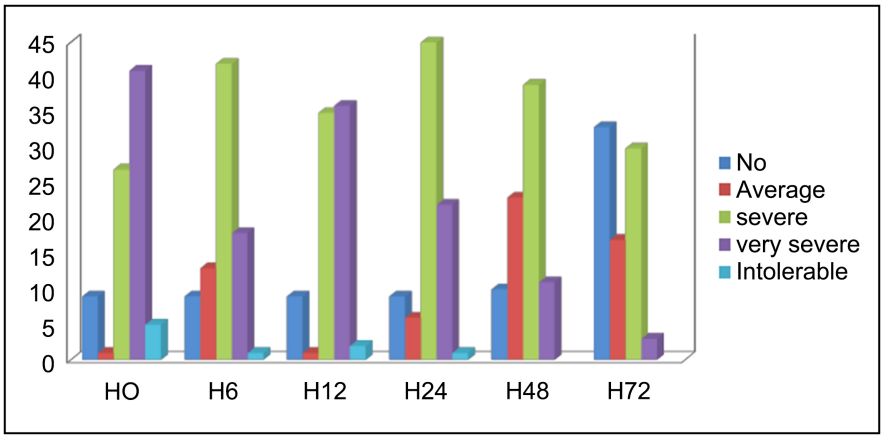

Figure 1. Distribution of patients by pain intensity.

Table 3. Distribution of patients by site of pain.

Site of pain	Workforce	Percentage
Upper limb	3	3.6
Thorax	4	4.8
Abdomen	4	4.8
Dorsal	2	2.4
Lower limb	16	1.3
Mixe = 2 seats and more	**45**	**54.2**
No	7	8.4
Undetermined	2	2.4
Total	**83**	**100**

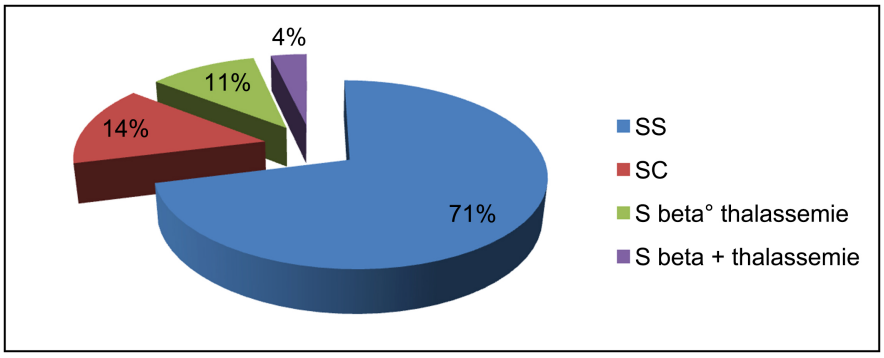

Figure 2. Distribution of patients by hemoglobin electrophoresis.

Lactated ringer and 0.9% saline were used together in 83.1% of cases. A level II analgesic was used in 87.95% of our patients. About 40% of our patients were relieved of their crisis in less than 3 days. The most commonly used antibiotic was third generation cephalosporin (26.3%) followed by cephalosporin + aminoside (25.3%). Red blood cells were transfused in 36.1% of children. Antimalarial drugs were used in 38.6%, oxygen in 28.9% and antipyretics in 60.24% of patients.

4. Discussion

4.1. Frequency

From 1 June 2017 to 31 May 2018, we collected 83 records of children with sickle cell crises out of 1777 children admitted to the pediatric emergency department, a frequency of 4.67%. This frequency is higher than that of Ouédraogo-Yugbaré S O [3] who obtained a frequency of 1.67% at CHUP-CDG and 0.22% at CMSC. The pediatric emergency department is the only structure to which children in emergency situations are referred and which ensures continuity of care over 24 hours.

4.2. Socio-Demographic Aspects

The age range of 60 to 120 months was the most frequent with 43.4% (n = 36). The average age was 94 months with extremes ranging from 8 to 180 months.

Our mean age is higher than that of Shongo M Y P [4] (39.1 months) because of the study population which was 6 months to 5 years in his study while it is 6 months to 15 years in our study.

In our study, the sex ratio was 1.4, close to the results of Cissouma A [5] in Mali and Abdala K A *et al.* [6] in the DRC who found a sex ratio of 1.4 and 1.5 respectively.

Dreux O [7] found an equal sex ratio, but Shongo M Y P *et al.* [4] in the DRC found a sex ratio of 0.7. These differences would be related to the demographics of each country.

For the occupation of the mothers, housewives were the most represented with 62.7%, of which more than half (53%) were not in school, compared to

43.4% for the fathers. Diakité A A [8] also found a predominance of housewives (80%). The low literacy rate of women in our countries explains this result, which is likely to reduce the understanding of parents in the context of therapeutic education.

We found that 24.1% of the children were born of a consanguineous marriage and more than half of the children (55.4%) had a history of familial sickle cell disease. Seck N [9] and Belala A *et al.* [10] found the consanguinity link in 51.3% and 45% of cases respectively. The notion of familial sickle cell disease was found by Diakité A A [8] in 27.1% of cases. The prevalence of hemoglobin S, combined with the practice of endogamy, explains the high proportions of familial sickle cell disease in our context.

4.3. Clinical and Paraclinical Aspects

The majority of children had a correct vaccination status according to the EPI, *i.e.* 89.2%. Seck N [9] and Ouakasse S [11] found respectively 87.4% and 60% of patients correctly vaccinated according to the extended vaccination programme. Our result could be explained by the fact that EPI vaccines are free in Mali. The most recalled non-EPV vaccines were the pneumococcal vaccine (25.3%) followed by the typhus vaccine (26.5%). These results are similar to those of Seck N [9] who found 25.1% for the pneumococcal vaccine, 44% against salmonella and 26.7% against meningococcal. The high cost of vaccines in relation to family income explains the low rate of vaccination coverage at this level.

More than half of the children had a history of seizures (54.22%). Vaso-occlusive seizures were the most frequent reason for hospitalization (22.9%). Our results are lower than those of Seck N [9] *et al.* who found 58.5% CVO. A history of hand-foot syndrome was found in 37.3% of patients. Shongo M Y P *et al.* [4] found that more than half (56.1%) of sickle cell patients in their series presented with dactilitis as their first attack. This result could be due to the fact that in their methodology the age range was 0 - 5 years. Whereas we used children between 6 months and 15 years of age, which increases the proportion of children who will be diagnosed through other symptoms

The main reason for consultation was vaso-occlusive crises with 50.6%. Our results are lower than those of Elie A D A [12] who observed 66.4% of painful seizures as the reason for admission. Two-thirds (67.5%) of the patients had a delay in weight and height. This could be due to the chronic anemia, the high frequency of diseases in sickle cell patients. More than half of the patients (60.2%) had a fever at entry. This is due to the extreme susceptibility of sickle cell patients to infection. The major signs were osteoarticular pain with 67.5%, followed by abdominal pain and mucocutaneous pallor with 45.8% and 39.8% respectively. Ouakasse S [11] found a predominance of osteoarticular pain (70%) and pallor (70%). In the study by Shongo M Y P *et al.* [4], jaundice was observed in more than 60% of cases. This difference could be due to the inclusion criteria because our study concerns children in crisis (in the emergency room), whereas Shongo M Y P [4] worked on all sickle cell patients even outside the critical

phase.

The pain was of the pulsatile type in 24.1% of cases and of the torsion type in 19.3% of cases. At H0 of admission, 41 (49.4%) of our patients had very severe pain. On the other hand, Diakité A A [8] found that at admission (H0) the majority of attacks were moderate, *i.e.* 62.9%, followed by intense attacks (37.1%). This difference is due to the fact that our study concerned children in emergency situations. The absence of pain went from 9 children (H0) to 33 children (H72), and severe pain went from 33 children (H0) to 3 children (H72). This is due to the effectiveness of the analgesics used. More than half (54.2%) of our patients had multiple sites of pain at admission. Our results are superior to those of Diakité A A [8] who found 38.04% of children with multiple sites of pain on admission. This difference is due to the severity of the clinical pictures and the intensity of pain in our study. More than half of our patients (53%) did not know why their attack occurred, physical effort (15.7%) and stress (14.5%) were the most common factors found. On the other hand, fever was the most frequent triggering factor in the study by Diakité A A [8] with 38.6%.

In our series, the SS form predominated with 71.1%. Seck N [9] and Elie A D A [12] also found a predominance of homozygous SS forms with 87% and 84% respectively. This is due to the high prevalence of hemoglobin S in the Sahelian zone. Hyperleukocytosis was noted in 74.69% of our patients, with an average of 23,807 leukocytes. Ouakasse S [11] found 80% of her patients had hyper leukocytosis with an average of 14,894. A hemoglobin level of less than 10 g/dl was found in 85.5% of patients with an average of 7.75 g/dl and extremes ranging from 2.43 g/dl to 12.5 g/dl. Our results are similar to those of Diakité A A [8] who found an average of 7.72 g/dl and extremes ranging from 3 to 10.2 g/dl. Ouakasse S [11] in Morocco found in her study an average of 6.14 g/dl, with extremes ranging from 4.4 g/dl to 9.2 g/dl. The low hemoglobin level is due to the short life span of red blood cells in sickle cell disease. Normocytosis was present in 45.8% of the patients coming in crisis. Among 10 patients studied, Ouakasse S [11] found that 6 had normocytic anemia (60%). Normocytic anemia is a characteristic of SS sickle cell disease. Anemia was normochromic in 84.4% of patients. Ouakasse S [11] had found a rate of CCMH ranging from 24% to 33.5%, with a mean equal to 30.26%. More than half of the patients (51.8%) had a normal platelet count. Ouakasse S [11] found that 60% of patients had normal platelet count. Of the 60.3% of patients who had a thick blood drop, more than half had a positive drop (38.6%). Elie A D A [12] obtained 41.41% of malaria as a reason for hospitalization. Mali being a malaria endemic area, malaria remains one of the triggers of sickle cell crises. Abdominal ultrasound revealed abnormalities in 6 of the 7 children who underwent the examination. Six cases of splenomegaly and two cases of hepatomegaly were noted. However, we did not record any cases of cholecystitis, which is one of the usual complications of sickle cell disease. In the study by Ouakasse S [11] 2 patients underwent an echo-abdominal examination which showed splenomegaly in both cases. The frequency of splenomegaly (6 cases out of 7) is due to the splenic sequestration observed in sickle

cell disease.

Chest radiography was abnormal in 20 of the 27 children who had it done. In the study by Ouakasse S [11], chest radiography was requested and performed in 5 patients studied, only one of whom showed a lung focus. This difference is explained by the size of her sample (10). Pulmonary infection is one of the most frequent causes of hospitalization in our study. It is often recognized as a trigger for CVO.

In 83.1% of children, hyperhydration with lactated ringer (LR) and saline (SS) 0.9% was done to amend the crisis. Abdala K A *et al.* [6] used hyperhydration (alone or combined with other treatments) in 71.6% of cases.These high rates of hyperhydration can be explained by the importance of hyperhydration in the management of CVO. A level II analgesic was used in 87.95% of our patients. Diakité AA [8] found that 87.1% of the pain was relieved thanks to level I analgesics and 12.9% which required switching to level II analgesics. Oukasse S [11] and Khelfaoui [13] in their studies found 90% of patients treated with level I analgesics.

This difference can be explained by our methodology which consisted in including only those patients who required emergency care and the need to reduce the time to sedation of pain.

The majority of children (74.62%) received a cephalosporin-based antibiotic treatment of 3 generation (26.5%), followed by the combination C3G + aminoside (25.3%). This use of antibiotics has been reported by other authors such as Khelfaoui [13] in 70% of cases. About 40% of our patients were relieved of their attacks during the first three days of hospitalization. The average length of hospitalization was 10 days with extremes ranging from 2 to 18 days. Our average length of hospitalization is higher than that of Diakité A A [8] (4 days) and that of Abdala K A [6] who had a hospital stay of 2 to 5 days in 71.6% of cases. This long average length of stay is explained by the severity of certain cases, which require treatment in the emergency room and then a move to the ordinary hospital ward.

5. Conclusion

The vaso-occlusive crisis remains the main reason for consultation in sickle cell children at Gabriel Toure University Hospital. The management in our context consists, in addition to the correction of the triggering factors, of hyperhydration and the administration of analgesics and blood products.

Conflicts of Interest

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

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