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Profile of Sickle Cell Disease Patients Admitted for Management of an Acute Complication in the Pediatrics Department of the Cocody University Hospital (CHU)

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Abstract

Introduction: Sickle-cell anemia is the source of numerous potentially fatal complications in the absence of adequate management. The aim was to contribute to improving the management of acute complications of sickle cell disease. **Methodology:** This was a retrospective descriptive study. It took place in the pediatrics department of the Cocoy University Hospital (CHU) from January 1 to December 31, 2022. We included all patients hospitalized for management of acute complications of sickle cell disease. **Results:** We identified 81 cases. The hospital prevalence of acute complications of sickle cell disease was 17.48%. The mean age was 6 years 5 months, with a sex ratio of 1.02. In 92.6% of cases, children were known to have sickle cell disease prior to hospitalization, and the mean age of discovery was 34.01 months ± 29.273. The patients' electrophoretic profile was dominated by the SS form in 45.7% of cases and SFA2 in 32.1%. Follow-up was irregular in 61.7% of cases. Anemia, fever, pain and respiratory difficulty were present in 80.3%, 69%, 67.9% and 32.1% of cases respectively. APS was diagnosed in 6 patients. Infectious complications were dominated by malaria (39.5%). The majority of our children received antibiotic therapy (76.5%). Surgical treatment was undertaken in 5 patients with osteoarticular infections. The outcome was favorable in 91.40% of cases, with 2 cases of death. Average hospital stay was 8.20 days. **Conclusion:** Improving the vital prognosis of children with sickle cell disease requires effective diagnosis and management of acute complications.

Keywords

Sickle Cell Disease, Acute Complications, Children, Prognosis

1. Introduction

Sickle cell disease is the most common genetic hemoglobin disorder in the world [1] [2]. It is characterized by the mutation of a hemoglobin beta chain gene located on chromosome 11, resulting in the formation of an abnormal hemoglobin, called hemoglobin S (HbS) [3]. According to the WHO, there are over 120 million sufferers worldwide, 2/3 of them in Sub-Saharan Africa, where the prevalence of sickle cell trait in the general population sometimes exceeds 30% [4]. In Côte d'Ivoire, 12% of the population are carriers of hemoglobin S [5]. Major forms of the disease are subject to serious and often fatal complications. In the absence of treatment, 25% - 50% of patients die before the age of 5, and only 5% - 10% reach adulthood [6]. Since the closure of the Yopougon University Hospital (CHU) in 2019, we have had a large flow of sickle cell patients admitted to the pediatric emergency department of the Cocody University Hospital. Most of these patients are hospitalized following an acute complication of sickle cell disease. Cocody University Hospital is a third-level public hospital in the city of Abidjan that has become a reference center for sickle cell disease because it houses a clinical and biological hematology department. Children with sickle cell disease are mostly hospitalized in the pediatric department following an acute complication of the disease. It seemed appropriate to conduct this study, the general aim of which was to describe the management of these complications with a view to improving the vital prognosis of these patients.

2. Methodology

This was a retrospective descriptive and analytical study conducted over a 12-month period from January 1 to December 31, 2022, in the medical pediatrics department of the Cocody University Hospital. We included all children aged up to 15 years with sickle cell disease and a known electrophoretic profile who had presented with an acute complication related to their condition. Patients with unworkable medical records were not included in the study. The verbal consent of the head of department was previously obtained. After authorization from the medical management, we took possession of the hospitalized patients' files. We used an anonymous survey form containing: A unique identifier per file, the clinical file number, open, closed, and multiple-choice questions but transcribed in a coded form to allow computerized exploitation. Data was collected on an individual survey form from hospitalization records. The variables studied were sociodemographic, clinical, therapeutic and evolutionary. Data was entered into Excel 2013 and analyzed using SPSS18 software. For qualitative variables, we used the calculation of proportions. For quantitative variables, we calculated the means, standard deviation, and extremes. For the

comparison of two qualitative variables, the chi-square or Fisher exact test was used. The significance threshold was 5%. Our study received the approval of the various managers of the Cocody University Hospital and was carried out on medical records consequently there was no need to obtain written or verbal consent.

Operational definition:

- Z-score: to assess nutritional status in patients aged from 1 month to 60 months.
- BMI: to assess nutritional status in patients aged over 60 months.
- Follow-up quality was ensured by compliance with consultations according to the form of sickle cell disease, effective use of disease-modifying therapy and an up-to-date vaccination schedule.
- Acute complications were all complications developing in an acute manner (<7 days) requiring emergency hospitalization. There are three main groups. They are either anemic, infectious or ischemic.

3. Results

3.1. Prevalence and Socio-Demographic Characteristics

Over the study period, we recorded 81 patients admitted for management of acute complications of sickle cell disease out of 455 admissions, representing a hospital prevalence of 17.48%. The mean age was 77.80 months (6 years 5 months), with extremes ranging from 7 months to 168 months (14 years). Children aged between 6 and 12 years (72 months and 144 months) accounted for 58% of cases. There was no gender predominance (sex ratio = 1.02).

3.2. Sickle Cell Status and Outpatient Follow-Up

In 92.6% of cases, the children were known to have sickle cell disease prior to hospitalization. Among these known sickle cell patients, the age of discovery was around 6 to 12 months in 33.33% of cases. The mean age of discovery was 34.01 ± 29.273 months, with extremes of 7 and 146 months. Anemia accounted for more than half of the circumstances of discovery (54.66%). The electrophoretic profile of patients was dominated successively by the SS form in 45.7% of cases, and SFA2 in 32.1%. Follow-up was irregular in 61.7% of cases. No treatment was in progress in 14.8% of cases. Expanded program of immunization vaccines were up to date in 43.2% of cases. Non-programme vaccines were not up to date in 49.4% of cases. Table 1 summarizes information on sickle cell status and history.

3.3. Clinical Data and Complications

Clinical signs included fever, pallor, difficulty breathing and pain in 69%, 80.3%, 32.1% and 67.9% of cases respectively. Malaria and IRAB were noted in 39.5% and 29.62% of cases respectively. Osteoarticular pain accounted for 48.15%. Acute chest syndrome (ACS) was observed in 6 cases (7.4%). With regard to anemic complications, 60 cases (74.1%) of acute hemolysis of infectious origin and 5 cases (6.17%) of acute splenic sequestration (AS) were noted. Clinical data and complications are summarized in Table 2.

Table 1. Distribution of patients according to sickle cell status and history.

	Numbers	Percentage
Known sickle cell disease (n = 81)		
Yes	75	92.6
No	6	7.4
Age of discovery (n = 75)		
[6 - 12 months]	25	33.3
[13 - 24 months]	18	24.0
>24 months	32	42.7
Circumstance of discovery (n = 75)		
Anemia	53	70.7
Pain	20	26.6
Systematic work-up	2	2.7
Electrophoretic profile (n = 81)		
SS	48	59.3
SC	3	3.7
SFA2	26	32.1
SAFA2	4	4.9
Follow-up		
Regular	8	9.9
Irregular	53	65.4
No follow up	20	24.7
Background treatment (n = 75)		
Acfol	2	2.7
Tanakan + acfol	54	72.0
Hydroxyurea + tanakan + acfol	1	1.3
Hydroxyurea + acfol	7	9.3
None	11	14.7
EPI vaccines* (n = 81)		
Up to date	35	43.2
Not up to date	46	56.8
Total	81	100.0
Non-EPI vaccine (n = 81)		
Up to date	5	6.2
Not up to date	76	93.8
Total	81	100.0

EPI: expanded program on immunization.

Table 2. Distribution of patients according to clinical data and complications.

	Numbers (n = 81)	Percentage
Clinical data		
Fever	56	69.0
Pallor	65	80.3
Respiratory difficulty	26	32.1
Pain	55	67.9
Malnutrition (SAM and MAM)	30	37.0
Complications		
Infectious complications		
Malaria	32	39.2
IRAB	24	29.62
Osteoarticular infection	6	7.4
Urinary tract infection	13	16.04
Sepsis	8	9.9
Cholecystitis	1	1.23
Salmonellosis	3	3.7
Vaso-occlusive complications		
Bone pain crisis	39	48.1
Acute chest syndrome	6	7.4
Anemic complications		
Acute splenic sequestration	5	6.17
Acute hemolysis	60	74.1

3.4. Treatment and Progression

Hyperhydration, blood transfusion and antibiotic therapy were used in 100%, 80.24% and 76.5% of cases respectively. Analgesics were used in 64.2% of patients. Surgical treatment was indicated in 5 patients (6.2%). Hydroxyurea was initiated during hospitalization in 3 patients (3.7%). Four patients (4.9%) received non-invasive ventilation (NIV). Hospital stay was between 1 and 7 days in 64.2% of cases. The average hospital stay was 8.20 days ± 5.76. Outcome was favorable in 91.4% of cases, with death in 2 patients (2.5%) (Table 3).

3.5. Risk Factors for Polytransfusion

Age, sex, awareness of sickle cell disease status, quality of follow-up, and type of disease-modifying therapy did not influence the number of transfusions. Hemoglobin level at admission did influence the number of transfusions (Table 4).

Table 3. Breakdown by treatment and outcome.

	Number (n = 81)	Percentage
Treatment undertaken		
Oxygen therapy	16	19.8
Analgesic	52	64.2
Transfusion with non-phenotyped blood	60	74.1
Transfusion with phenotyped blood	05	6.17
Hyperhydratation	81	100
Antibiotic therapy	62	76.5
Anti-malarial	32	39.5
Hydroxyurea	3	3.7
NIV	4	4.9
Surgical treatment	5	6.2
Length of hospital stay in days		
<7	52	64.2
[8 - 14]	19	23.46
>14	10	12.35
Progression		
Exeat	74	91.4
Transfer	3	3.7
SCAM	2	2.5
Death	2	2.5

Table 4. Distribution according to risk factors for polytransfusion.

	Number of transfusions				p
	None	1 time	2 times	≥3 times	
Age					
<2 years	3 (75.0%)	1 (25.0%)	0 (0.0%)	0 (0.0%)	0.256
[2 - 5 years]	10 (38.5%)	5 (33.3%)	4 (15.4%)	5 (19.2%)	
[6 - 12 years]	9 (19.1%)	4 (19.0%)	6 (12.8%)	21 (44.7%)	
≥13 years	1 (25.0%)	1 (25.0%)	1 (25.0%)	1 (25.0%)	
Sexe					
Boy	12 (29.3%)	8 (19.5%)	7 (17.1%)	14 (34.1%)	0.640
Female	11 (27.5%)	12 (30.0%)	4 (10.0%)	13 (32.5%)	

Continued

Knowledge of sickle cell status					
Yes	19 (25.3%)	19 (25.3%)	11 (14.7%)	26 (34.7%)	0.177
No	4 (66.7%)	1 (16.7%)	0 (0.0%)	1 (16.7%)	
Follow up quality					
Good	2 (50.0%)	1 (25.0%)	0 (0.0%)	1 (25.0%)	0.502
Irregular	13 (26.0%)	11 (22.0%)	7 (14.0%)	19 (38.0%)	
Bad	4 (19.0%)	7 (33.3%)	4 (19.0%)	6 (28.6%)	
Unknown	4 (66.7%)	1 (16.7%)	0 (0.0%)	1 (16.7%)	
Basic treatment					
Tanakan	0 (0.0%)	0 (0.0%)	0 (0.0%)	1 (100.0%)	0.136
Acfol	2 (100.0%)	0 (0.0%)	0 (0.0%)	0 (0.0%)	
Tanakan + acfol	16 (27.6%)	17 (29.3%)	9 (15.5%)	16 (27.6%)	
Hydroxyurée + acfol	0 (0.0%)	0 (0.0%)	1 (14.3%)	6 (85.7%)	
None	5 (41.7%)	3 (25.0%)	1 (8.3%)	3 (25.0%)	
Hemoglobin level in G/DL					
<3	1 (9.1%)	5 (45.5%)	2 (18.2%)	3 (27.3%)	0.004
[3 - 5[	3 (10.3%)	5 (17.2%)	3 (10.3%)	18 (62.1%)	
[5 - 7[	8 (36.4%)	7 (31.8%)	4 (18.2%)	3 (13.6%)	
[7 - 10]	10 (58.8%)	2 (11.8%)	2 (11.8%)	3 (17.6%)	
>10	1 (50.0%)	1 (50.0%)	0 (0.0%)	0 (0.0%)	

4. Discussion

4.1. Prevalence and Socio-Demographic Characteristics

The hospital prevalence of acute complications of sickle cell disease was 17.80%. This prevalence is much higher than that of Alain. F [6] in Madagascar, who found a prevalence of 2.2%. This high prevalence of sickle-cell disease in our series is thought to be due to the closure of the Yopougon University Hospital (reference center for hematological pathologies), which led to patients being redirected to the Cocody University Hospital. The 6 - 12 age group was the most affected, *i.e.* 58%, with an average age of 6 years 5 months, as found by several authors such as Alain. F [6] and Dah Sassan in Côte d'Ivoire [7]. In the literature, the recurrence of hyper-algesic vaso-occlusive crises is very often seen from the age of 7 [6]. We have not noted any gender predominance. In literature. Ibrahim keita [8] and Alain F. [6] found a male predominance, with sex ratios of 1.4 and 1.3 respectively. In contrast, Kpakoutou N. A. [9] in Mali found a female predominance in 68.88% of cases. This

difference reflects the autosomal nature of sickle cell disease transmission.

4.2. Sickle Cell Status and Outpatient Follow-Up

Sickle cell disease was discovered during hospitalization in 6 children (7.4%). The mean age of discovery was 34.01 months ± 29.273 with extremes of 7 and 146 months. Ibrahim Keita [8] found in his series that 86.1% of cases were diagnosed during hospitalization, and the age range of discovery was 24 to 59 months. These results show the delay in screening for sickle cell disease in our countries, often in the face of complications. On the other hand, in Europe and specifically in France, screening is carried out systematically in the neonatal phase [10]. The SS form was found in the majority (45.7%), followed by the SFA2 form (32.1%), which is in line with the studies by Ibrahim Keita [8] and Akolly. D [11], who reported 41% and 84% homozygous forms respectively. The frequency of SS and Sβ phenotypes in our study can be explained by the frequency of heterozygous AS phenotypes and the coexistence of β-thalassemia in the Mediterranean region, as well as the continued existence of consanguineous marriages in our country [12].

Follow-up was not regular in 61.7% of cases. Studies of sickle-cell anemia in the African context note irregular follow-up of patients. Akolly. D [11] found irregular follow-up in 68% of cases. As sickle cell disease is a chronic pathology for which curative treatment is currently unavailable, improving prognosis requires regular follow-up to prevent life-threatening complications [13]. Infections in sickle-cell patients could be prevented by vaccination against certain germs. In our series, as in many others, vaccination coverage was low [8]. This could be explained by ignorance, neglect and lack of awareness of the need to vaccinate sickle-cell children correctly, and also by financial difficulties, especially for vaccines not covered by the Expanded Programme on Immunization. The absence of satisfactory vaccination coverage in these sickle-cell children is a source of serious complications.

4.3. Clinical Data and Complications

Anemia, fever, pain and respiratory difficulty were present in 80.3%, 69%, 67.9% and 32.1% of cases respectively. Barry IK [14] found fever in 100% of cases. The near-presence of fever could be explained by the extreme susceptibility of sickle cell patients to infection [14]. The pallor of the anemia could be explained by the hemolytic nature of sickle cell disease. The majority of patients (71.6%) had good nutritional status, and only 4.9% were undernourished. In contrast, Akolly. D [11] found malnutrition in 36.36% of cases. Malnutrition is more frequent in children with sickle cell disease and must be systematically detected and managed through regular follow-up, as it aggravates the various morbid conditions [14].

Infectious complications were dominated by malaria (39.5%) and pneumonia (29.64%). Akolly. D [11] reported 41.41% cases of malaria. Malaria was the leading cause of hospitalization in both our study and that of Akolly. D [11]. Barry IK [14] ranked pulmonary infections 1st (44.1%). According to the literature, pulmonary infections are the leading cause of hospitalization in children with sickle cell dis-

ease [15] [16]. Indeed, the functional asplenia present in patients with major sickle cell syndrome, and particularly in SS or S/β-thalassemic forms, results in increased susceptibility to encapsulated germs (S.pneumoniae, N.meningitidis, H. influenzae). These patients are therefore at high risk of bacterial infection [17] [18]. Vaso-occlusive complications were dominated by painful osteoarticular crises (48.1%) and APS (7.4%), a serious, life-threatening complication. Acute hemolysis due to malaria or bacterial infection was found in 74.1% of patients.

4.4. Therapeutic and Evolutionary Data

Antibiotic therapy is the treatment of choice for hospitalized sickle cell patients, and was indicated in 76.5% of cases. According to the literature, when infection is suspected, it is recommended to start antibiotic treatment empirically, without waiting for bacteriological culture results [19]. Transfusion was carried out in 80.24% of children, mainly with non-phenotyped blood, due to the difficult access to phenotyped blood in our emergency situation. Only 3 patients were started on hydroxyurea during hospitalization, and 4 patients were already on hydroxyurea. Hydroxyurea is recommended only for severe forms of sickle cell disease in children over 2 years of age [19]. Morphine is an important drug in the management of CVO in patients with sickle cell disease. Morphine is not yet widely used by prescribers [20], due to a lack of availability and, above all, its difficult handling, which requires a suitable device and rigorous monitoring [21].

The average length of hospital stay was 8.20 days, comparable to that of Kpakoutou [9], who found 7.5 days. This long hospital stay increases the burden of care for these sickle-cell patients, whose parents have to cope with the already high costs of monitoring their sickle-cell-affected children. The outcome was favorable in 91.40% of cases, which could be explained by the department's experience in caring for sickle-cell children. Akolly. D [11] also found a favorable outcome in 95.31% of cases. Two patients (2.5%) died of severe acute anemia. This result is lower than that of Kpakoutou [9], who reported 4.44% deaths.

5. Conclusion

Sickle-cell anemia is the source of many potentially fatal acute complications in the absence of effective management. Management consists essentially in preventing complications, hence the importance of regular multidisciplinary medical follow-up, with paediatricians playing an important role. Improving the vital prognosis of the sickle-cell child requires effective diagnosis and management of acute complications.

Conflicts of Interest

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

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