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Sickle Cell Disease in Children at Agadez (Niger): Epidemiology, Diagnosis and Management

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Abstract

Objective: To determine the epidemiological, diagnostic, and therapeutic aspects of major sickle cell syndromes followed at the Agadez Mother and Child Health Center. **Materials and Methods:** This was a prospective, descriptive, and analytical study including all sickle cell patients, regardless of age or sex, registered from February 1, 2025, to July 31, 2025. **Results:** 150 cases of sickle cell disease were recorded out of 2578 patients, representing a frequency of 5.81%. The majority of patients were between 24 and 60 months old (34%). Males predominated, with a male-to-female ratio of 1.5. Consanguinity was reported in 40% of cases. The homozygous SS sickle cell diseases was predominant, accounting for 99.33%. Hand-foot syndrome was the most frequent presenting symptom, occurring in 32.67% of cases. Acute hemolytic anemia was the most frequent acute complication, occurring in 95.33% of cases. Anemia was severe in 39.33% of cases. All patients received at least one therapeutic education session. Treatment was primarily preventive, focusing on avoiding the triggering factor. **Conclusion:** Sickle cell disease remains a public health problem in Niger and requires improved diagnostic and therapeutic resources.

Keywords

Sickle Cell Disease, CSME, Agadez, Niger

1. Introduction

Sickle cell disease is a hemolytic anemia that causes red blood cells to sickle, leading to biochemical disturbances and varied clinical manifestations depending on age and the type of major sickle cell syndrome [1]. In Africa, 150,000 to 300,000 children are born with sickle cell disease each year, of whom 60% to 80% die before the age of five due to inadequate management [2]. Sickle cell disease represents a major public health issue in Niger due to its high prevalence, estimated at 4.2% [3]. It is often discovered in the context of infections, hemolysis, or vaso-occlusive crises [4]. Its course is marked by complications that can be acute or chronic [5]. Treatment has been revolutionized by hydroxyurea [6]. The lack of data on this condition in the Agadez region motivated this study at the Mother and Child Health Center in this region, aiming to investigate the epidemiological, diagnostic, and therapeutic aspects of major sickle cell syndromes at this center.

2. Materials and Methods

- Study Type and Sample

This was a retrospective, descriptive study conducted from February 1, 2025, to July 31, 2025.

- Inclusion Criteria

All sickle cell patients, regardless of age or sex, diagnosed via hemoglobin electrophoresis and followed at the Zinder Mother and Child Health Center, who provided consent.

- Diagnostic and Therapeutic Methods

Sickle cell disease was diagnosed based on clinical suspicion, abnormal blood count findings, or systematic screening. Our diagnoses were confirmed by hemoglobin electrophoresis at alkaline Ph. Follow-up and management of sickle cell patients were carried out by two pediatricians and general practitioners. Diagnostic and therapeutic costs were borne by the patients, regardless of the number or type of complications. This study was conducted within this context.

- Statistical Analysis

The collected information included age, sex, sociodemographic data, clinical manifestations, and diagnostic and therapeutic approaches. Data were analyzed using Epi Info software version 7.2.6.0. Pearson's chi-squared test was used to examine relationships between qualitative variables. The test was considered significant if the p-value was <0.05.

- Study Limitations

The extremely low socioeconomic status of parents resulted in the non-performance of certain follow-up tests and the unavailability of specific therapeutic agents in the Agadez region.

3. Results (Table 1)

3.1. Sociodemographic Data

Frequency

Table 1. Recapitulative results.

Variables		Number	Percentage (%)
Gender	Male	90	60
	Female	60	40
Age (months)	0 to 24	33	22
	24 to 60	51	34
	60 to 120	37	24.6
	Above 120	29	19.4
Phenotype	SS	149	99.33
	SC	1	0.67
Discovery Circumstance	Hand-foot syndrome	49	32.67
	Sucking	46	30.67
	CVC	41	27.33
	Fever	33	23.33
	Systematic	15	10
	Jaundice	11	7.33
Acute Complications	Hemolysis	143	95.33
	Infection	119	79.33
	CVC	109	70.33
	Stroke	6	4.00
	STA	3	2.00
Chronic Complications	Osteonecrosis	0	-
	Retinopathy	0	-
	Cardiac Complications	0	-
Hemoglobin Level	≤6	68	45.33
	6 to 9	59	39.33
	≥10	14	9.33

During the study period, 150 cases of sickle cell syndrome were identified among 2570 patients, representing a frequency of 5.81%. Among these, 99.33% (n = 149) were homozygous SS and 0.67% (n = 1) were SC.

Age and Sex

Male patients were predominant at 60% (n = 90), while female patients accounted for 40% (n = 60), resulting in a male-to-female sex ratio of 1.5. The majority of patients fell within the age group of 24 to 60 months, representing 34% (n = 51). The mean age was 63.15 months, with extremes ranging from 2 months to 192 months.

Geographic Origin and Occupation

Most patients resided in the Urban Commune of Agadez, accounting for 80.67% (n = 121) of cases. The majority of fathers of sickle cell children were unschooled, representing 53.34% (n = 80), with most of them being merchants (53.34%, n = 80). Mothers were unschooled in 82.67% (n = 124) of cases, with the majority being homemakers (82.67%, n = 124).

Personal History

First and second consanguinity was observed in 40% (n = 60) of cases. More than half of the patients had received one to five blood transfusions, accounting for 61.33% (n = 92) of cases. Additionally, 49.33% (n = 74) of patients had up-to-date EPI vaccination status

3.2. Clinical Data

The majority of sickle cell syndromes were diagnosed before 24 months of age in 67.33% (n = 101) of cases, with a mean age at diagnosis of 18.33 months. Hand-foot syndrome (Figure 1) was the most common presenting feature, observed in 32.67% (n = 49) of cases.

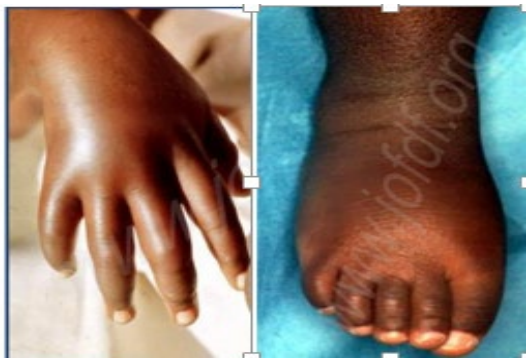

Figure 1. Hand-foot syndrome.

Followed by pale skin and mucous membranes in 30.67% (n = 46) (Figure 2).

Figure 2. Pale skin and mucous membranes.

Vaso-occlusive crises in 27.33% (n = 41), fever in 23.33% (n = 35), and jaundice in 7.33% (n = 11) of cases (Figure 3).

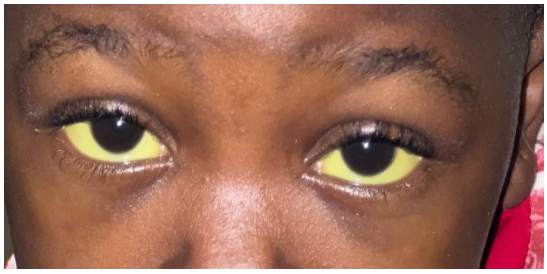

Figure 3. Jaundice.

Acute complications were dominated by acute drop in hemoglobin levels in 95.33% (n = 143) of cases, followed by infections in 79.33% (n = 119) and bone vaso-occlusive crises in 64.67% (n = 97). No chronic complications were recorded.

3.3. Paraclinical Data

The majority of sickle cell syndromes, 99.33% (n = 149), were homozygous SS, while 0.67% (n = 1) were SC. Most patients had hemoglobin levels between 6 and 9 g/dL in 39.33% (n = 59) of cases. The anemia was normochromic normocytic in 36% (n = 54) of cases. Severe anemia (hemoglobin ≤ 6 g/dL) was observed in 45.33% (n = 68) of patients. Blood group O Rh-positive was the most common, accounting for 36% (n = 54) of cases.

3.4. Therapeutic Data

All the children's parents received therapeutic education, representing 100% of cases. Patients who were regularly followed up accounted for 90% (n = 135) of cases. Children under 5 years of age were systematically placed on penicillin V.

4. Discussion

4.1. Epidemiological Data

During the study period, 150 cases of sickle cell syndrome were identified among 2570 patients, representing a frequency of 5.81%. Djibrilla-Almoustapha A *et al.* in Niger (Diffa in 2024 and Dosso in 2025) reported frequencies of 4.88% and 1.08%, respectively [7] [8]. Among the cases, 99.33% were homozygous SS. Cissouma A *et al.* in Mali reported a frequency of 57% for the SS form [9]. The majority of patients fell within the age group of 24 to 60 months, accounting for 34% (n = 51). The mean age was 63.15 months, with extremes ranging from 2 months to 192 months. Ngolet LO *et al.* in Brazzaville, Mali, reported a mean age of 26.04 years [10]. Male patients were predominant at 60%, with a sex ratio of 1.5. Sonia Douamba *et al.* in Burkina Faso in 2017 reported 51.9% in favor of females [11]. Djibrilla-Almoustapha A *et al.* in Niger in 2021 found a male predominance with a sex ratio of 1.3 [12]. This difference is likely due to chance, as sickle cell disease is an autosomal recessive disorder. The majority of fathers of sickle cell children were unschooled (53.34%), and merchants accounted for 53.34% of cases. Mothers were unschooled in 82.67% of cases, with the majority being homemakers

(82.67%). Elhadji-Chefou M *et al.* in Niger (Maradi) in 2024 reported similar findings [13]. These factors may impact the quality of care. Consanguinity was observed in 40% of cases. Diallo D in Mali in 2016 reported 32.8% of cases [14]. However, this result is lower than that of Nehoulne G in Chad in 2003, who reported 52% of cases [15].

4.2. Clinical Data

The majority of sickle cell syndromes were diagnosed before 24 months of age in 67.33% of cases, with a mean age at diagnosis of 18.33 months. In Niger in 2024, Abba Kaka HY *et al.* (2025) and Inoussa BD *et al.* (2022) reported similar findings [16] [17]. Hand-foot syndrome was the most common presenting feature, observed in 32.67% of cases. Djibrilla-Almoustapha A *et al.* in Niger (Diffa) in 2024 reported similar results [7]. Acute complications were dominated by acute drops in hemoglobin levels (95.33%), followed by infections (79.33%). Diarra Y *et al.* in Burkina Faso reported a predominance of infectious complications [18].

4.3. Paraclinical Data

Most patients had hemoglobin levels between 6 and 9 g/dL (39.33%). The anemia was normochromic normocytic in 36% of cases. Severe anemia was observed in 45.33% (n = 68) of patients. Djibrilla-Almoustapha A *et al.* in Niger (Diffa and Dosso) reported similar findings [7] [8].

4.4. Therapeutic Data

All patients received therapeutic education (100%). This is an important step in the management of sickle cell disease, aimed at preventing significant complications. Patients who were regularly followed up accounted for 90% of cases. Diarra Y in Burkina Faso reported 80% of cases with regular follow-up [18].

Of the patients, 49.33% (n = 74) had up-to-date EPI vaccination status. Sonia Douamba *et al.* in Burkina Faso reported 81.2% [11]. This low vaccination coverage rate may be linked to purchasing power on one hand and/or irregular follow-up on the other, both associated with low educational levels.

5. Conclusion

Sickle cell disease is a genetic condition that presents numerous diagnostic and preventive challenges. It remains a public health issue in Niger.

Ethical Aspects

This publication complied with ethical and professional standards, in particular the protection of patient identity and their approval regarding the use of images for educational purposes.

Conflicts of Interest

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

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