

Factors Associated with Non-Adherence to Treatment in Sickle Cell Patients Monitored at the National Reference Center for Sickle Cell Disease in Niger

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Abstract

Introduction: Sickle cell disease is a real public health problem in the world and particularly in Niger where the prevalence of the S gene is estimated at 25% and that of the homozygous forms at between 1% and 2%. Treatment combines quarterly follow-up of patients and management of complications. The objective of this study was to identify the potential explanatory factors of non-adherence to treatment in sickle cell patients followed at the national reference center for sickle cell disease in Niger. **Methods:** This is a cross-sectional study of sickle cell cases followed at the CNRD in Niger. The population consisted of all sickle cell patients followed in this center in 2021. The data collection techniques were individual interviews and documentary reviews. Non-adherence was assessed with the Girerd test. Descriptive statistical tests and simple and multiple logistic regression models were performed. **Results:** A total of 368 patients were enrolled. The median age is 7 years (4; 10) and the sex ratio is 1.04. Ninety-eight (98) or 26.6% were compliant and 270 (73.4%) were non-compliant. In multivariate analysis, the factors independently and negatively associated with non-adherence to treatment were schooling (adjusted OR [95% CI], p-value), 0.17 [0.10 - 0.30]; p < 0.0001; discontinuation of treatment [OR = 0.15 [0.05 - 0.43], p < 0.0004; the distance from the Center 0.58 [0.33 - 0.99], p ≤ 0.0489. Unawareness of the disease was positively and significantly associated with non-adherence 7.68 [2.20 - 26.72], p ≤

0.0014. **Conclusion:** The factors influencing treatment compliance identified in this study are all modifiable. To prevent the complications of sickle cell disease, we must fight against ignorance, make care services accessible and make care free.

Keywords

Associated Factors, Sickle Cell Disease, Non-Adherence to Treatment, Niger

1. Introduction

Sickle cell disease is one of the most important hemoglobinopathies (thalassaemia, sickle cell disease, etc.) in the world in terms of frequency (with approximately 50 million people affected) and social impact, recently recognized as a global public health problem by the World Health Organization (WHO) [1].

The management of sickle cell disease like any chronic disease is lifelong and requires good compliance. To ensure this good compliance requires both clinical and psychosocial care.

Indeed, therapeutic compliance refers to the concordance between the behavior of the patient vis-à-vis his treatment and the recommendations of his doctor [2].

With regard to compliance with medication prescriptions, 30% to 60% of patients with a chronic disease including sickle cell disease can be categorized as being poorly or non-compliant. Several factors can influence this parameter. The characteristics of the patient, the particularities of the disease, the methods of treatment, the attitudes of the doctor or the organization of the healthcare system are the main incriminated factors [2]. Other factors can also be considered, in particular the psychological dimension of the treatment, which we will not discuss in this study.

As a result, compliance with treatments prescribed for chronic diseases represents a major public health issue worldwide. In addition, in its latest report, the World Health Organization highlighted that poor adherence to long-term treatments is a growing problem [3].

Thus, several studies have been carried out on therapeutic compliance in the world. Some of them have found that there is an association between adherence and the main demographic, medical and psychosocial correlates others have found that the association depends on the patient's age [4] [5] [6].

In Niger so far, no study has focused on treatment compliance in sickle cell patients in order to determine the factors associated with it. This study is however necessary to explain the level of therapeutic observance and to research the factors associated with this observance.

The objective of this study was to identify the potential explanatory factors of non-adherence to treatment in sickle cell patients followed up at the national reference center for sickle cell disease in Niger.

2. Methods

2.1. Type of Study

This is an analytical cross-sectional study conducted among sickle cell patients monitored at the national reference center for sickle cell disease in Niger. The collection was made from June 14 to July 24, 2021, *i.e.* a period of forty (40) days. Informed consent from patients was always required and the study did not expose patients to additional risks.

2.2. Study Population

The population consisted of all patients with major sickle cell syndrome who are followed at the national reference center for sickle cell disease in 2021.

Prop nt included in our study patients aged 18 and over with more than one year of follow-up at the center and patients aged under 18 with more than one year of follow-up at the center whose parent or caregiver in charge gave his consent to participate in this work.

The only non-inclusion criterion was a no target, but who interrupted monitoring at the CNRD for more than 3 months

The sample size was calculated on the basis of the prevalence (P) of non-adherence estimated at 64.74% [7]. The sample size of the primary targets is defined by OpenEpi[®]: $n = 368$.

2.3. Data Collection

The survey took place over a period of 40 days from June 14 to July 24, 2021. Patients meeting the inclusion criteria were recruited during the follow-up consultation for sickle cell patients. The collection of information was done through a questionnaire completed by the investigator according to the answers of the patient or his tutor and the information of his follow-up sheet for certain data. The questionnaire had two parts; the first part included the identification of the patient, the socio-demographic characteristics (age, sex, marital status of the parents, number of children with sickle cell disease in the family, place of residence, educational level of the parents, socio-economic level of the parents, status at with regard to social security), the therapeutic characteristics (specific features of the disease, treatment methods, organization of the healthcare system, knowledge of the disease). The second part was made up of questions relating to the assessment of therapeutic compliance, taken from the Girerd questionnaire, which consists of 6 questions. The questions take into account the aspects of regularity in taking medication, discontinuation of treatment, forgetfulness and the quantity of medication to be taken.

Assessment of the GIRERD test score

Total yes	Assessment of the level of compliance
0	Good compliance
1 - 2	Minimal compliance issue
3	Poor compliance

This questionnaire was administered in the local dialect, and tested beforehand with 10 patients to detect any comprehension problems.

2.4. Analysis Plan

At the end of the inclusion period, the data was entered into Excel software with prior coding of the various responses and then analyzed using EPI info and SPSS software.

2.5. Descriptive Study

First, a descriptive analysis was performed by calculating the confidence interval (95% CI) around a percentage for a risk = 0.05. There is a 5% risk that the percentage obtained will be outside this interval.

2.6. Study of Factors Associated with Non-Adherence to Treatment

The second part of the study concerns the identification of factors associated with non-adherence to treatment. Two groups of patients were formed for this purpose: the group of non-compliant patients and the group of compliant patients. In the univariate analysis, the associations between the dependent variable (therapeutic non-adherence) and all of its determinants were tested by the Chi2 test or Fisher's exact test when the Chi2 validity conditions were not fulfilled. A p-value of less than 0.05 was considered statistically significant. Double contingency 2×2 cross tables were established for the calculation of the odds ratio (OR) as an epidemiological association factor with calculation of the confidence intervals (95% CI) of the risk. A multivariate analysis by logistical regression was carried out by choosing as dependent variable the binary variable "non-compliance" with two modalities: non-observant and observing. The explanatory variables included in the multivariate model are those for which the association with the dependent variable has a statistical significance level below 0.20 during the univariate analysis. Multivariate analysis was used to calculate the adjusted odds ratios and their confidence intervals for each of the factors studied. The explanatory variables included in the multivariate model are those for which the association with the dependent variable has a statistical significance level below 0.20 during the univariate analysis. Multivariate analysis was used to calculate the adjusted odds ratios and their confidence intervals for each of the factors studied. The explanatory variables included in the multivariate model are those for which the association with the dependent variable has a statistical significance level below 0.20 during the univariate analysis. Multivariate analysis was used to calculate the adjusted odds ratios and their confidence intervals for each of the factors studied.

2.7. Description of the Variables to Be Studied

The non-compliant patient is the one with poor compliance or a minimal compliance problem, *i.e.* having a yes score greater than or equal to 1, and the pa-

tient who is therapeutically compliant is the one with good compliance.

Sociodemographic characteristics (age, sex, marital status of parents, number of children in the family, place of residence, level of education of parents, socioeconomic level of parents, social security status) and perception to be informed about sickle cell disease were informed on the declaration of the patients. Therapeutic characteristics (particularities of the disease, treatment methods, organization of the healthcare system, knowledge of the disease) were obtained from declarations by patients confronted with the content of the follow-up sheets.

3. Results

3.1. Epidemiological Characteristics of Patients

A total of 368 patients were investigated, 51% of whom were female, the sex ratio was 1.04F/H, the median age was 7 years (4.10), 92.1% had the SS form, the majority of patients resided in Niamey, *i.e.* 79.1%. Two hundred and thirty-eight (238) or 64.6% of the parents surveyed are educated and 35.3% of them had no level of education, 206 of the parents worked in the private sector or 55.9%.

Three hundred and eight (308) parents of the children, or 83.7%, lived as a couple, 61.4% of whom were polygamous. The family's monthly income is more than 100,000 CFA francs in 60.9% of cases and 286 or 77.7 did not have health insurance (**Table 1**).

3.2. Clinical Features

Specifics about the disease

Among the patients, 61.7% had known their status for 5 years, 63.6% had less than 5 attacks per year and 91.3% had been hospitalized less than 3 times a year.

Modality of treatment

Compared to treatment, 14.9% of sickle cell patients had stopped their treatment, 3.8% thought that there were no risks associated with stopping treatment, 24 or 43.6% mentioned traditional treatment as due to discontinuation of their medical treatment. 197 or 53.5% of sickle cell patients took only 2 tablets per day and 278 or 75.5% performed less than 5 examinations per year.

Knowledge about the disease

Three hundred and twenty-three (323) or 87.8% had knowledge of their disease, 92.7% of them had been informed by their doctor, 64.7% mentioned forgetting as a reason for not knowing the disease (**Table 2**).

System organization

50.8% of sickle cell patients were located less than 10 kilometers from the care center, only 76 or 20.6 used their own personal means of transport to get to the center, 35.6% admitted to having experienced a drug shortage linked to a lack of resources in 79.3% of cases (**Table 2**).

Level of Therapeutic compliance in sickle cell patients

It emerges from this distribution that the patients are compliant in 26.6% of cases and the non-compliant represent 73.4% (**Figure 1**).

Table 1. Socio-demographic and clinical characteristics of sickle cell patients followed.

Socio-demographic and clinical characteristics		Number, n	Percentage (%)
Sex	F	188	51
	M	180	49
Place of residence	Other regions	76	20.6
	Niamey	292	79.3
Schooling	No	130	35.3
	Yes	238	64.7
Educational level	Primary	47	12.8
	Primary	100	27.1
	Superior	91	24.7
	No Level	130	35.3
Parents' occupations	Without	59	16
	Private sector	206	56
	Public sector (civil servants)	103	28
Marital status	Couple	308	83.7
	Separated	52	14.1
	Widow	8	2.2
Matrimonial regime	Monogamy	189	61.4
	Polygamy	119	38.6
Monthly income	<100,000	144	39.1
	≥100,000	224	60.9
Health Insurance	No	286	77.7
	Yes	82	22.3
Form of sickle cell disease	CS	29	7.9
	SS	339	92.1
Total		368	100

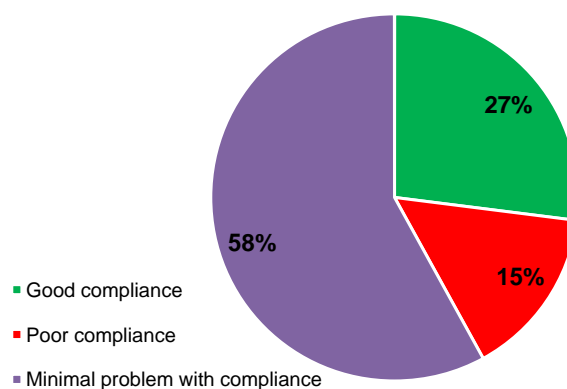
**Figure 1.** Distribution of sickle cell patients according to the level of therapeutic compliance.

Table 2. Distribution of patients according to knowledge about sickle cell disease & health system organization.

Knowledge about the disease & health system organization		Number, n	Percentage (%)
Knowledge of the disease	No	45	12.2
	Yes	323	87.8
Person who informed the patient of his illness	Other practitioner	27	7.3
	Doctor	341	92.7
Reasons for the ignorance of the disease	Lack of information	4	11.8
	Oversight	22	64.7
	No information	8	23.5
Reasons for drugs shortage	Lack of means	79	61.7
	Neglect	5	3.9
	Out of stock	44	34.4
Distance from center	<10 KM	187	50.8
	≥10 KM	181	49.2
Means of transport used (staff)	No	292	79.3
	Yes	76	20.7
Break in drugs	No	237	64.4
	Yes	131	35.6
Total		368	100

Particularity of the disease in sickle cell patients

Among the patients, 61.68% had known their status for 5 years, 63.59% had less than 5 attacks per year and 91.30% had been hospitalized less than 3 times a year (Table 3).

Factors associated with non-adherence to treatment in sickle cell patients: univariate analysis

The univariate analysis shows that the socio-demographic and clinical factors which influence compliance with treatment are: age OR = 0.56 (0.33 - 0.95), $p = 0.032$, residence OR = 2.50 (1.26 - 4.98) $p = 0.007$, health insurance OR = 0.44 (0.23 - 0.84) $p = 0.012$, schooling OR = 0.19 (0.11 - 0.32) $p < 0.001$, distance from the center OR = 0.38 (0.23 - 0.62) $p < 0.0001$, Stopping treatment OR = 0.29 (0.16 - 0.51) $p = 0.0001$ and the number of analyzes requested per year OR = 0.33 (0.17 - 0.63) $p = 0.0006$ (Table 4).

Multivariate analysis

The multivariate analysis showed that there is a significant association between non-adherence to treatment and the following factors: Stopping treatment OR = 0.15 (0.05 - 0.43) $p = 0.0004$, knowledge of the disease OR = 7.68 (2.20 - 26.72) $p = 0.0014$, distance from the health center OR = 0.58 (0.33 - 0.99) $p = 0.04$, schooling OR = 0.17 (0.10 - 0.30) $p = 0.0001$ and Health Insurance OR = 0.36 (0.16 - 0.84) $p = 0.01$ (Table 5).

Table 3. Distribution of patients according to the particularity of the disease in sickle cell patients at the national reference center for sickle cell disease of Niamey in 2021.

Specifics about the disease		Number, n	Percentage (%)
Duration of sickle cell status	<5 years	141	38.3
	≥5 years	227	61.7
Number of crises per year	<5	234	63.6
	≥5	134	36.4
Number of hospitalizations per year	≤3	336	91.3
	>3	32	8.7
Total		368	100

Table 4. Distribution according to treatment in sickle cell patients at the national reference center for sickle cell disease modalities in Niamey in 2021.

Processing methods		Number, n	Percentage (%)
Discontinuation of treatment	No	313	85
	Yes	55	15
Existence of risk when stopping treatment	No	14	3.8
	Yes	354	96.2
Number of drugs taken per day	≤2	197	53.5
	>2	171	46.5
Reasons for stopping treatment	Lack of means	17	30.9
	Neglect	9	16.3
	Oversight	5	9.1
	Traditional treatment	24	43.6
Number of analyzes requested per year	Without	59	16
	≤5	278	75.5
	>5	90	24.5
Total		368	100

Table 5. Socio-demographic and clinical characteristics of sickle cell patients in univariate and multivariate analyses.

Variables	Observation level		ORb (95% CI)	p-value	ORa (95% CI)	p-value
	Watching	Non-observant				
Age range	<5 years	68	216	1		
	≥5 years	30	54	0.56 (0.33 - 0.95)	0.032	0.96 (0.46 - 1.97)
Residence	Niamey	87	205	1		
	Other regions	11	65	2.50 (1.26 - 4.98)	0.007	1.81 (0.66 - 4.98)
Sex	M	44	136	1		
	F	54	134	0.80 (0.50 - 1.27)	0.35	
Monthly income	>100,000	59	165	1		
	≥100,000	39	105	0.9725 (0.6 - 1.54)	0.87	

Continued

Health Insurance	Yes	13	69	1				
	No	85	201	0.44 (0.23 - 0.84)	0.012	0.36 (0.16 - 0.84)	0.01	
Diet	Monogamy	53	136	1				
	Polygamy	77	231	1.54 (0.8 - 2.67)	0.12	1.45 (0.74 - 2.82)	0.27	
Schooling	Yes	36	202	1				
	No	98	270	0.19 (0.11 - 0.32)	<0.001	0.17 (0.10 - 0.30)	0.0001	
Knowledge of the disease	Yes	95	233	1				
	No	3	37	5.02 (1.51 - 16.70)	0.0037	7.68 (2.20 - 26.72)	0.0014	
Distance from center	<10 km	33	153	1				
	≥10 km	65	117	0.38 (0.23 - 0.62)	<0.0001	0.58 (0.33 - 0.99)	0.04	
Number of children with sickle cell disease in the family	≤ 2	7	39	1				
	>2	91	231	0.45 (0.19 - 1.05)	0.06	0.98 (0.35 - 2.76)	0.98	
Occupation	Without			1				0.007
	Private sector			1.93 (0.96 - 3.87)				
	Public sector			0.82 (0.36 - 1.86)				
Educational level	No level			1				<0.001
	Primary			0.10 (0.03 - 0.30)				
	Primary			0.30 (0.17 - 0.55)				
	Superior			0.13 (0.06 - 0.29)				
Number of hospitalizations/year	≤3	9	23	1				
	>3	89	247	1.08 (0.48 - 2.43)	0.84			
Number of crises per year	≤5	21	86	1				
	>5	77	184	0.58 (0.33 - 1.00)	0.05	0.70 (0.32 - 1.53)	0.38	
Number of drugs per day	≤ 2	55	142	1				
	>2	43	128	1.15 (0.72 - 1.83)	0.0014	1.91 (0.90 - 4.06)	0.09	
Use of personal vehicle	Yes	18	117	1				
	No	80	153	0.74 (0.41 - 1.31)	0.30			
Stopping treatment	Yes	18	117	1				
	Boop	80	153	0.29 (0.16 - 0.51)	0.0001	0.15 (0.05 - 0.43)	0.0004	
Form of sickle cell disease	SS	91	7	1				
	CS	248	22	1.15 (0.47 - 2.79)	0.75			
Number of analyzes requested per year	<5	12	80	1				
	≥5	86	190	0.33 (0.17 - 0.63)	0.0006	1.06 (0.42 - 2.70)	0.88	
Duration of sickle cell status	<5 years	46	52	1				
	≥5 years	181	89	0.43 (0.27 - 0.69)	<0.001	0.51 (0.24 - 1.10)	0.088	

ORb: Gross odds ratio. ORa: Adjusted odds ratio.

4. Discussion

4.1. Therapeutic Compliance

In our study, 73.37% were non-compliant and 26.63% had good compliance. Oudin Doglioni *et al.* [8] found that 74.4% of respondents had poor compliance, 24.2% average compliance and 1.4% good compliance [8], Candrilli *et al.* [7] found non-adhesion at 64.7% in their study. As Adewoyin *et al.* in a study carried out in Nigeria which finds a high proportion of non-adherence 80% [9]. This could be explained on the one hand by the cost of taking charge of the disease and on the other hand by the chronic nature of the disease. The more the disease becomes chronic, the more the financial resources are exhausted, the more the patience of the parents to support the attendance of the care centers is reduced.

4.2. Factors Significantly Associated with Non-Adherence to Treatment in Sickle Cell Patients

We had found a significant association between non-adherence and the level of schooling OR = 0.17 with $p < 0.0001$. This could be explained by the fact that 64.67% of the children's parents were educated. The more parents are educated, the more they know about the disease and its socioeconomic and health consequences.

We also found a significant association between knowledge of the disease and non-adherence to treatment (ORa = 7.68, $p = 0.0014$). In this study 87.77% of the parents knew about the disease of their children. According to Lainé, 71.4% know their diseases in West Africa and 83.3% in Central Africa [10], the more worrying the disease becomes, the more parents seek to know it better.

This study also showed an association between stopping treatment and non-adherence to treatment (ORa = 0.15, $p = 0.0004$). Stopping the treatment causes serious complications in the patients suddenly, when the parents return to the center they become more observant for fear of inflicting the same suffering on the child.

Finally, we found a significant association between non-compliance and the distance from the treatment center (ORa = 0.58, $p = 0.04$). The closer the center, the easier the accessibility and above all that in the study more than three quarters (3/4) of the patients had no means of personal transport, which made accessibility difficult. In this study only 20.65% of the patients come from other regions, which testify to the inadequacy in the accessibility to care.

Our study had limitations in the methodology and type of study. Indeed, the methodology for evaluating therapeutic compliance used is that proposed and validated by Girerd. This Girerd questionnaire has not been validated in sub-Saharan populations and not translated into language, we had to make an approximate translation in order to adapt it to our realities. While our study has highlighted certain factors that influence treatment compliance, a larger cohort study over a longer period would certainly provide more reliable information on the management of this chronic condition, sickle cell disease.

5. Conclusions

At the end of this study, it turns out that non-adherence to treatment in sickle cell patients is a frequent and multifactorial phenomenon. Therapeutic non-adherence is associated with factors such as knowledge of the disease, discontinuation of medical treatment, distance from the care center and level of schooling. In view of our results and to improve this observance, we must necessarily fight against ignorance, and make sickle cell treatment services accessible, make treatment free, because one third of people who have stopped treatment had done for lack of means.

Identifying the factors associated with non-adherence allows governments and practitioners to identify bottlenecks and intervene to improve adherence. Therapeutic education and psychosocial support are necessary to motivate the patient to face this disease.

6. Limit of Study

Our study allowed a punctual evaluation of the therapeutic observance in patients followed at the national reference center for sickle cell disease in Niamey. Although analytical cross-sectional, our study made it possible to include about twenty variables in the study and to identify certain factors associated with non-compliance with treatment in sickle cell patients.

However, it has limitations:

- ✓ Indeed, an important variable (ethnicity) is not entered in this study. The fact that the ethnic data were not collected while we know the conservation of genetic defects in the frequency/presence of sickle cell disease.
- ✓ The methodology for evaluating therapeutic compliance used is that proposed and validated by Girerd. This Girerd survey has not been validated in sub-Saharan populations and not translated into language, we had to make an approximate translation in order to adapt it to our realities.
- ✓ If our study has highlighted certain factors that influence treatment compliance, a study of a larger cohort over a longer period would certainly provide more reliable information on the management of this chronic condition that is sickle cell disease.

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Conflicts of Interest

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

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Appendix

Questionnaire

I) Socio-demographic and epidemiological characteristics

Age []

Age < 5 years [] >5 years []

Gender M [] F []

Place of residence: Niamey other regions to be specified.....

Parents' profession: civil servant [] private [] without []

Health insurance yes [] No []

Marital status: [] Couple [] Widowed [] separated []

Regime: Monogamy [] Polygamy []

Education: Yes [] No []

If yes level: Primary [] Secondary [] Higher []

Monthly household income: <XOF 100000 [] >XOF100000 []

What form of sickle cell disease do you have? SS [] SC [], S b thalassemia []

Number of children with sickle cell disease in the family.....

II) Particularity of the disease

- How long have you known your sickle cell status < 5 years [] >5 years []

How many hospitalizations per year? ...

How many crises per year?

III) Method of treatment

Are you currently on treatment? yes no

How many medications do you take? (Number)

Have you ever stopped treatment once? Yes [] No []

If yes, why?

Do you think there is a risk in stopping your treatment?

Have you had any tests for your illness: Yes [] No []

How many times have you been asked for tests?

How many times have you made them?

Why have you not been carried out all the analyzes:

- High Cost [] Oblivion [] Distant Lab []

- Lack of information [] Other [] specify .

IV) Knowledge about the disease

- Do you know your disease? Yes [] No []

If not why ? Never got information [] Forgot [] Doesn't care []

Who told you about your illness? Your doctor [] Another practitioner []

Other:

V) Organization of the system

Distance from the center: <10 km [] >10 km []

Means of transport used: Personal Yes [] no []

Have you ever had any drug shortages? yes [] No []

If yes, why? lack of means [] Out of stock at the pharmacy [] others.....

VI) GIRED compliance assessment test

Did you forget to take your medicine this morning? YES NO

Since the last visit, have you run out of medication? YES NO

Have you ever taken your treatment later than usual?

YES NO

Have you ever not taken your treatment because some days your memory fails you? YES NO

Have you ever skipped your medication because some days you felt like your medication was doing you more harm than good? YES NO

Do you think you have too many tablets to take? YES NO