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Assessment of Cognitive Disorders and Their Impact on School Performance in Children and Adolescents with Sickle Cell Disease at the Albert Royer Children's Hospital, Dakar, Senegal

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Abstract

Introduction: Sickle cell disease can cause chronic cerebral arteriopathy with numerous complications, including neurocognitive disorders. In Africa, there are few studies on neurocognitive disorders. The objective of our study was to determine the cognitive disorders observed in children with sickle cell disease and their impact on their schooling. **Patients and Methods:** This was a cross-sectional, descriptive study with an analytical focus conducted between May 14, 2021, and July 31, 2021 (2 months and 17 days) among SS sickle cell children aged 6 to 16 years who were regularly monitored in the USAD outpatient clinic. They were matched with healthy children of the same age and sex. The Wechsler scale, WISC-V, was used for the neuropsychological assessment of the children. **Results:** Twenty-four children were included, with a mean age of 11.62 years and extremes of 6 and 16 years. A male predominance was noted (54.16%). All cognitive functions were affected, with working memory and visuospatial abilities being the most severely impaired. In terms of schooling, 13 children had repeated at least one year, representing a repetition rate of 54.16%, and the dropout rate was 50%. **Conclusion:** Sickle cell disease is a common condition in Senegal with numerous neurological complications, particularly neurocognitive deficits. These deficits can have a negative impact on

children's education and socialization.

Keywords

Sickle Cell Disease, Neurocognitive Deficits, Children, Senegal

1. Introduction

Sickle cell disease is a genetic disorder of hemoglobin that is transmitted in an autosomal recessive manner and is very common in sub-Saharan Africa [1]. It is a systemic disease and its complications can affect all organs. There are numerous neurological complications, most often affecting the central nervous system (stroke, epilepsy, cognitive impairment, and psycho-behavioral disorders) and rarely the peripheral nervous system [2]. These complications are disabling both motor and psychologically, with a negative impact on the socialization and schooling of these children. On our continent, while there are many studies on strokes related to sickle cell disease, there is little data on neurocognitive disorders [3] [4]. In Senegal, a few studies on neurological complications related to sickle cell disease have been conducted, but none of them address cognitive disorders related to sickle cell disease. It is in this context that we conducted this study, the aim of which is to assess the consequences of sickle cell disease on the neurocognitive development of children and their schooling.

2. Materials and Methods

We conducted a prospective, cross-sectional, descriptive study for analytical purposes during the period from 14 May to 31 July 2021 (2 months and 17 days). Our study population consisted of children aged 6 to 16 years old being treated for SS sickle cell disease. They were paired with non-sickle cell (healthy) pupils from primary and secondary schools in the Jaaraf Ibra Faye complex in Dakar-Plateau, of the same age, educational level and socio-economic status. This work was carried out at three different sites:

- The Neuropsychology Unit and the Outpatient Care Unit for Children and Adolescents with Sickle Cell Disease, located within the Albert Royer National Children's Hospital (CHNEAR) in Dakar.
- The Jaaraf Ibra Faye complex, consisting of an elementary school and a middle school (CEM) under the supervision of the Dakar-Plateau Education and Training Inspectorate (IEF). We carried out neuropsychological assessments of the control children with the authorisation of the administrative and parental authorities.

Our study included all SS sickle cell children aged between 6 and 16 who were regularly monitored as outpatients at the USAD. Not included in our study were all SS sickle cell children being treated for psychiatric disorders (depression, mania, schizophrenia, etc.), taking psychotropic drugs (anxiolytics, hypnotics, mood

stabilisers, neuroleptics, etc.), or presenting with motor deficits (dysarthria, aphasia, epilepsy, motor and sensory deficits, and balance disorders) or children whose parents or guardians did not agree to participate in the study.

We used randomised and reasoned sampling. Interviews with patients were conducted on consultation days (Monday to Friday) and patients were chosen on a first-come, first-served basis. We systematically took the last patient on the list, due to the length of the interviews and in order not to compromise the order in which patients were seen by the paediatrician. Healthy children were selected after the sick children had been tested in order to meet the matching criteria, namely the same age, gender and educational level. The Wechsler scale (WISC V), adapted to our socio-cultural context (some unfamiliar examples and questions were contextualised by using examples more familiar to children), was used. The intelligence quotient (IQ), calculated on the basis of seven subtests (similarities, vocabulary, cubes, matrices, balances, digit memory, codes) and the main indices were used for each child in order to gain an insight into cognitive disorders. However, optional or additional tests were not used. The total duration of the session varied between 1 hour and 1 hour 30 minutes. One of the guardians attended the session but remained in the background. The Gaussian normal curve was used to read the IQ and the various main indices of the WISC-V.

3. Results

We collected data on 24 children with sickle cell disease, with an average age of 11.62 ± 2.49 years and ages ranging from 6 to 16 years. The most represented age group was 6 to 12 years (Table 1). In our study, there was a predominance of males, accounting for 54.16% (13 boys). All the children in our study population were enrolled in public schools, with French as the official language, at different levels of education. Sixteen children were enrolled in primary school and eight in secondary school (two in 6th, 5th, 4th and 3rd grades). The academic difficulties encountered by children with sickle cell disease were:

- Repeating a year: 13 children had repeated at least one year during their schooling, representing a repeat rate of 54.16%. Among them, six children (46.15%) had repeated twice, four children (30.76%) had repeated three times and 23.07% had repeated once.
- School dropout: 12 children had dropped out of school, mainly due to absenteeism (10 children), poor academic performance (12 children) and repeating a year (12 children).

Neuropsychological assessment of children with sickle cell disease revealed impairment in all areas of cognition compared to controls (see Figure 1). Based on the indices, we found the following: Intelligence quotient: The mean total score for children with sickle cell disease was 49.95 ± 9.7, with extremes ranging from 40 to 76. According to the Gaussian normal curve, 23 children with sickle cell disease had a very low IQ, *i.e.* 95.83%. For the control group, the mean was 88.72 ± 4.12 with extremes of 80 and 97.

Table 1. Distribution of patients by age group.

Age groups (years)	Number	Proportions (%)
[6 - 12]	16	66.67
]12 - 16]	8	33.33
Totals	24	100

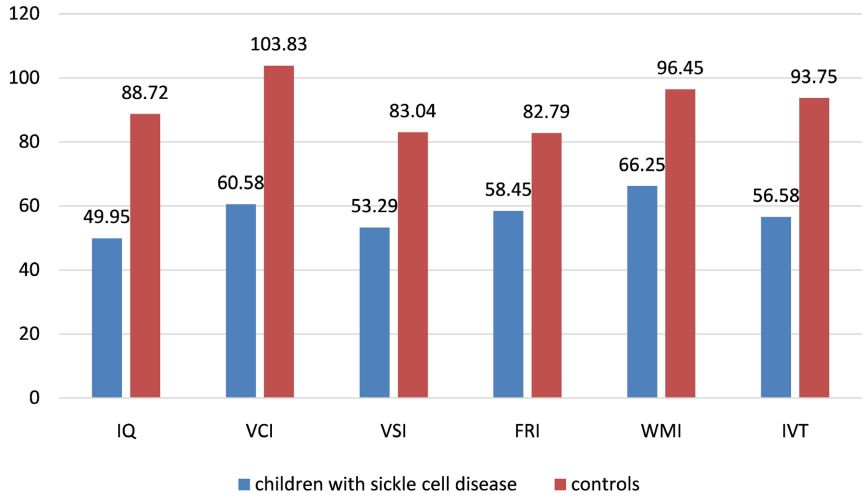

Figure 1. Comparison of average IQs and principal indices between children with sickle cell disease and controls.

Verbal comprehension index (VCI): the mean was 60.58 ± 15.48 with extremes of 45 to 89, and 16 children had a very low VCI. The mean total score was 103.83 ± 4.74 with extremes of 98 and 113 for the controls.

Visuospatial Index (VSI): 23 children had a very low VSI with a mean of 53.29 ± 7.85 and extremes ranging from 45 to 72. In the controls, the mean VSI was 83.04 ± 2.94 with extremes ranging from 78 to 89.

Fluid reasoning index (FRI): the mean total score was 58.45 ± 8.17 with extremes ranging from 45 to 76, and 22 children had a very low FRI. The mean FRI was 82.79 ± 5.25 with extremes ranging from 76 to 91 in the controls.

Working memory index (WMI): the mean was 66.25 ± 9.30 with extremes ranging from 51 to 88. According to the Gaussian normal curve, 16 children had a very low WMI. The mean WMI for the controls was 96.45 ± 6.39 with extremes ranging from 82 to 107.

Processing speed index (PSI): 22 children had a very low PSI, and the mean total score was 56.58 ± 17.43 with extremes ranging from 45 to 126. The mean PSI for the controls was 93.75 ± 7.63 with extremes ranging from 75 to 105.

In our study, we found certain factors that were significantly associated with neurocognitive disorders, namely anaemia, haemoglobin S and foetal haemoglobin levels.

Anaemia:

All children with sickle cell disease had anaemia, but to varying degrees. Sixteen

children had moderate anaemia (haemoglobin levels between 6 and 8) and eight children had mild anaemia (haemoglobin levels between 8 and 11). No children had severe anaemia. Children with sickle cell disease and mild anaemia had IQs ranging from 50 to 76, with an average of 60.25, while those with moderate anaemia had an average IQ of 49.31, ranging from 40 to 60. Patients with mild anaemia had better scores on the following indices: ICV, IRF, IMT, IVT (see Table 2).

Table 2. Distribution of main indices according to the degree of anaemia in children with sickle cell disease.

Degree of anaemia	The main indices of the WISC-V				
	VCI	VSI	FRI	WMI	PSI
Mild anaemia	60.88	53	59.76	66	58.58
Moderate anaemia	59.85	54	55.28	66.85	51.71

Foetal haemoglobin level:

For children with a foetal haemoglobin level below 10, the average IQ was 45.6, with extremes of 40 and 50. For those with a foetal haemoglobin level between 10 and 20, the average IQ was 48.4. The average IQ was 55.5 for children with sickle cell disease and a foetal haemoglobin level above 20 (see Figure 2). Patients with high foetal haemoglobin levels had better scores on the following indices: ICV, IVS, IRF, IMT, IVT (see Table 3).

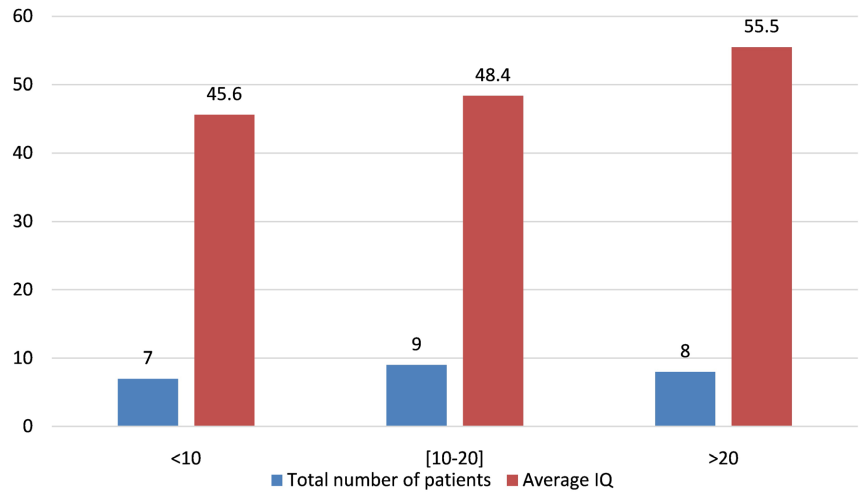

Figure 2. Distribution of average IQ scores according to foetal haemoglobin levels in children with sickle cell disease.

Table 3. Relationship between principal indices and foetal haemoglobin levels in individuals with sickle cell disease.

Hb F level	VCI	VSI	FRI	WMI	PSI
≤10	50.28	49	56.71	61.14	51.57
10 - 20	60.22	54.55	58.66	67.88	51.44
>20	70	55.62	59.75	68.87	66.75

Haemoglobin S level:

In our study, the average IQ was 44 for patients with haemoglobin S levels above 90, and the average IQ was 50 for children with haemoglobin S levels between 70 and 90. For sickle cell patients with haemoglobin S levels below 70, the average IQ was 58.5 (see Figure 3). Patients with low haemoglobin S levels had better scores on the following indices: ICV, IVS, IRF, IMT, IVT (see Table 4).

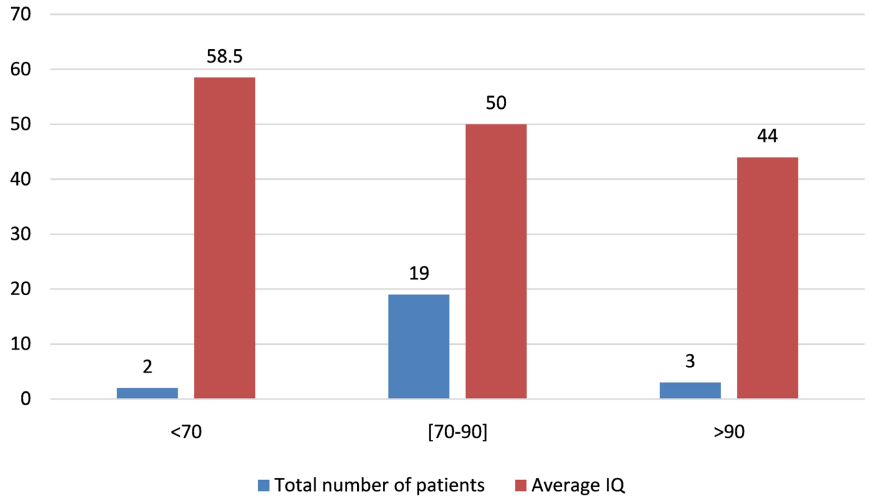

Figure 3. Distribution of IQs according to haemoglobin S levels in sickle cell patients.

Table 4. Relationship between principal indices and haemoglobin S levels.

HbS	VCI	VSI	FRI	WMI	PSI
<70	77	60.5	58	74	59
70 - 90	61.31	53.63	59.15	66.21	56.36
>90	45	46.3	54.33	61.33	56.33

4. Discussion

In Africa, there is little data on the frequency of cognitive disorders in children with sickle cell disease [3] [4]. In our study, the average age was 11.62 years, which contradicted the data in the literature, where cognitive deficits affected adolescents more [5] [6]. A male predominance was observed in our study, at 54.16%. It should be noted that many studies did not find a gender predominance [4] [7]. Sickle cell disease is a chronic cerebral arteriopathy and can be responsible for deficits affecting various areas of cognition, namely general intelligence, attention, executive functions, memory, language, visuomotor skills and academic performance [8]. In studies of children with sickle cell disease with or without documented brain damage, all cognitive functions were often affected, particularly in the study by Steen, R. G *et al.* (2005), which highlighted impaired performance on total IQ measures, which was more severe in children with sickle cell disease and cerebral ischaemia.

Language functions are also affected in cases of sickle cell disease. According to

Brown *et al.* [9], children who have had a clinical stroke make more errors in a naming task than children with no abnormalities on MRI. However, Schatz *et al.* [10] observed no significant difference in language measures between children with and without silent strokes. Armstrong *et al.* [11] found impaired written language in children with sickle cell disease who had had a silent stroke, compared to children without infarction. In our study, we noted impaired language functions in 16 children, or 66.66%.

Deficits in visuospatial abilities were noted in all children with sickle cell disease in our study. Our results were similar to data in the literature, where we noted impaired visuomotor skills in children with sickle cell disease. However, some authors, such as Armstrong *et al.* [11], found more impaired performance in children with sickle cell disease who had a clinical stroke than in children with a silent stroke or no brain damage. In contrast, Brown *et al.* [9] found no difference between children with clinical and silent strokes.

At school, the sickle cell children in our study had difficulties, with an estimated repetition rate of 54.16% and a dropout rate of 50%. Wang *et al.* [12] showed that children with sickle cell disease who had suffered a stroke performed less well than children with sickle cell disease who had no abnormalities on MRI. Schatz *et al.* [10] found that children with sickle cell disease who had silent strokes had twice as many academic difficulties as children with sickle cell disease without neurological abnormalities. They also found that 58% of children with silent strokes repeated a year or received special education, compared to 27% of those without brain abnormalities. [10]. It is essential to identify silent strokes as early as possible, as they have serious consequences on cognitive functioning, academic performance and the health of children with sickle cell disease. The use of a neuropsychological battery would then make it possible to identify the presence of significant cognitive deficits and refer these subjects for further medical evaluation [9].

Patients in our cohort had moderate anaemia (70%) and mild anaemia (30%). The negative effect of anaemia affected all areas of cognition and was more severe in sickle cell patients with moderate anaemia compared to those with mild anaemia. These results are consistent with the expected effects of chronic cerebral hypoxia on cognition [13].

In our study, sickle cell patients with high foetal haemoglobin levels performed better in cognitive domains. There is a link between foetal haemoglobin and cognitive functioning. These results are interesting in our context, where hydroxyurea is one of the therapeutic options indicated in certain situations. The desired effect is to stimulate the production of foetal haemoglobin. In a preliminary study, Puffer *et al.* [14] compared the cognitive performance of a group of sickle cell patients treated with hydroxyurea for more than a year (n = 15) and a group of sickle cell patients who had never been treated with hydroxyurea (n = 50). The authors observed better performance in the group of patients on hydroxyurea in the areas of verbal comprehension, fluid reasoning and general intellectual ability. These results corroborate the positive impact of foetal haemoglobin on cognitive function.

5. Conclusion

Neurological disorders are common in children with sickle cell disease, with numerous complications that can be life-threatening and affect the children's functional and, above all, cognitive abilities. In our study, brain MRI allowed us to establish a radiological-clinical correlation and possibly establish neuropsychological testing as a standard diagnostic tool for silent strokes, especially in countries where MRI is expensive and inaccessible. Neuropsychological assessment is an important tool for facilitating the social and educational adaptation of children with sickle cell disease and should be carried out systematically in children with sickle cell disease.

Conflicts of Interest

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

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