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Transcranial doppler echo in children with sickle cell SS in Niger: Velocimetric study

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Abstract
Aim: The aim of our work is to study the velocities by trans cranial Doppler in children with homozygous sickle cell SS, more specifically to measure cerebral circulatory velocities by trans cranial oppler in children with sickle cell SS and determine the relationships between the results of trans cranial Doppler with the age and type of sickle cell disease.
Materials and Methods: We conducted a retrospective prospective study (October 2018 to December 2020) and over a period of 27 months.
Results: We recruited a total of 126 patients aged between 1 year and 15 years. The sex ratio is 1.2. The average systolic velocities at the level of the right and left middle cerebral artery are abnormally high respectively with a rate of 61.90% and 58.73%. In total we had 68.25% (n=86) of patients with high risk, 9.52% (n=12) with intermediate risk.
Conclusion: Sickle cell disease is one of the most common genetic pathologies in our regions, especially the homozygous form. The latter is the one that presents more complications and among which we have Stroke. Stroke detection is done using transcranial Doppler, hence the purpose of our study.

Keywords: SS sickle cell disease, trans cranial doppler, Niger

Introduction

Hemoglobinopathies are hereditary abnormalities of hemoglobin and sickle cell disease represents the most widespread defect in the world with 100 million individuals affected. It is estimated that more than 300,000 children are born each year worldwide, mostly in low- and middle-income countries, with a severe form of hemoglobinopathy (SS). Approximately 5% of the world population are asymptomatic carriers of a sickle cell or thalassemia gene; this percentage reaches 25% to 40% in certain regions [1].

It is a real public health problem in Niger due to its prevalence on the one hand, its complications (anemic, ischemic and infectious) leading to repeated and prolonged hospitalizations as well as the mortality it causes on the other hand [2]. One of the most feared complications is stroke, this complication affects 6 to 17% of children and young adults worldwide [3]. The risk of stroke is higher in the first decade and especially in SS sickle cell patients [3, 4].

Thus, transcranial Doppler (TCD) is a non-invasive exploration technique of the intracranial arteries. Since its first use by Aasliden 1982, its indications have continued to expand. Its use in children with sickle cell disease stems from the work of Adams who in 1992 showed that children with high velocities in the middle cerebral or internal carotid arteries had an increased risk of stroke [5].

In developed countries, the detection of high-risk patients coupled with a regular transfusion program has led to an exponential reduction in strokes [6]. Africa is home to the largest number of sickle cell patients in the world but struggles to implement policies to prevent complications [7].

In Niger, screening for cerebral vasculopathy by transcranial Doppler is not systematic; therefore, there are no data on the velocimetric study of sickle cell patients.

So it seemed useful to us to carry out this study whose objective is to measure cerebral circulatory speeds by DTC in children with SS sickle cell disease and determine the

relationships between the results of DTC with age and sex.

Materials and Methods

We conducted a retrospective prospective study (October 2018 to December 2020) over a period of 27 months.

We included in this study children with SS homozygous sickle cell disease, aged less than or equal to 15 years, received for transcranial Doppler ultrasound.

The transcranial Doppler examination was carried out with mindray brand devices, either a DC70 or a DC80. In all cases the examination was carried out by the same operator. A 2-4 MHZ probe was used. The examinations were carried out using the temporal window (right, left) and the occipital window using color Doppler and pulsed Doppler. The average maximum speeds, obtained after an automatic tracing of the envelope of a cycle, were recorded on the anterior, middle, posterior cerebral arteries and the basilar trunk. Thus after recording the results, the risk of cerebral vasculopathy is established according to the following classification: the risk is low when the average systolic speed is less than 170 cm/s; the risk is said to be intermediate when the average systolic speed is between 170 and 200 cm/s; the risk is said to be high when the average systolic speed is greater than or equal to 200 cm/s

We studied the following parameters.

- Age
- Sex
- Reasons for the examination

Parameters explored using transcranial ultrasound

- Average systolic velocity of the right middle cerebral artery.
- Average systolic velocity of the left middle cerebral artery.
- Average systolic velocity of the right anterior cerebral artery.
- Average systolic velocity of the left anterior cerebral artery.
- Average systolic velocity of the right posterior cerebral artery.
- Average systolic velocity of the left posterior cerebral artery.
- Average systolic velocity of the basilar trunk.

The data were recruited using Excel 2013 software. The analysis of the data obtained was carried out with Epi info 7.2 software with a significance threshold of less than 0.05.

Results

We included 126 patients in our study. The average age of our patients is 6.67±3.51 years. The age group of 1-10 years is the most present with a percentage of 85.72% (table 1). The male and female gender are approximately equal with 49.21% and 50.79% respectively. The most frequent reason for examination is the follow-up assessment with a percentage of 61.9% (Table 2). On transcranial Doppler at the level of the right and left anterior cerebral artery 34.13% and 30.95% of patients have a VSM ≥ 200 cm/s (figure N°1), at the level of the right and left middle cerebral artery respectively 61.90% and 58.73% have a VSM ≥ 200 cm/s (figure N°2) , at the level of the posterior artery 21.43% of patients on the right and left have a VSM ≥ 200 cm/s and at the level of the basilar trunk 33.33% have a VSM ≥ 200 cm/s.

The risk of vasculopathy was high in 68.25%, intermediate in 9.52% of cases and low in 14.29% of cases (Table 3).

Table 1: Distribution of patients by age group.

Age range (Years)	Frequency	Percent
[1-5]	51	40, 48
[6-10]	57	45, 24
[11-15]	18	14, 29
Total	126	100, 00

Table 2: Distribution of patients according to age groups by indication for transcranial Doppler.

	Age range (Years)			
Indication	**[11-15]**	**[1-5]**	**[6-10]**	**Total**
History of transient ischemic attack	0	1	0	1
History of stroke	1	4	3	8
Monitored assessment	10	37	28	75
Headache	5	2	9	16
Control	2	5	11	18
Troubleshooting	0	1	2	3
After-effects of stroke	0	1	3	4
Chronic vertigo	0	0	1	1
Total	18	51	57	126

Discussion

The WHO reports 120 million people carrying the sickle cell mutation. Sickle cell disease is one of the most common genetic diseases in the world and 300,000 new SS sickle cell patients are born each year worldwide, with more than 200,000 cases recorded in Africa [8].

For the African continent, the WHO indicates a prevalence of 13%. In West African countries such as Ghana and Nigeria, the frequency of the trait reaches 15 to 30% according to the WHO [9]. The prevalence of sickle cell trait according to the 2010 WHO report is around 25% in Niger, Niamey being an area located in the sickle cell belt [10].

During its evolution several manifestations can appear among which we have cerebral manifestations, the most feared of which is stroke. Thus, sickle cell disease increases the risk of stroke in children by 220 [11].

The average age of our patients is 6.67 years ±3.51 and the age group of 1 to 10 years is the most present with a frequency of 108 cases or 85.72%. This frequency in the first decade can be explained by the fact that sickle cell disease is a genetic disease that is most often detected early in life [9, 10].

The homozygous SS type is the form of sickle cell disease most present in Niger, especially in West Africa [10]. It represents the form with the most complications, especially cerebral complications [7, 3, 5]. The interest of our work lies in the fact that our study population consists only of the homozygous SS form. In order to detect or diagnose cerebral vasculopathy, the most requested test is transcranial Doppler because it is a reliable and accessible test in our regions. It emerged from our study that the most frequent reasons for examination are the follow-up assessment in 61.9% of cases (n = 78) and the control in 14.29% of cases (n = 18). In terms of the above, we unfortunately note among our patients that the follow-up by the DTC is insufficient as is the control too. This observation made in our study goes against the recommendations made in the literature which highlights the importance of examining DTC in the follow-up of children with sickle cell disease aged between 2 and 15 years [3, 12].

The age group of 1 to 10 years is the one presenting more patients with high VSM, we find 58.73% of patients in this age group out of the 68.25% having a high risk. Speeds are highest between ages 3 and 12 with a peak speed around 7-8 years. It is interesting to note that this age group is that of maximum risk of stroke. Cerebral vasculopathy is more common in the first decade of life in sickle cell patients [12, 13]. This is why it is recommended to do transcranial Doppler in the annual follow-up of sickle cell patients, especially those aged between 2 and 15 years.

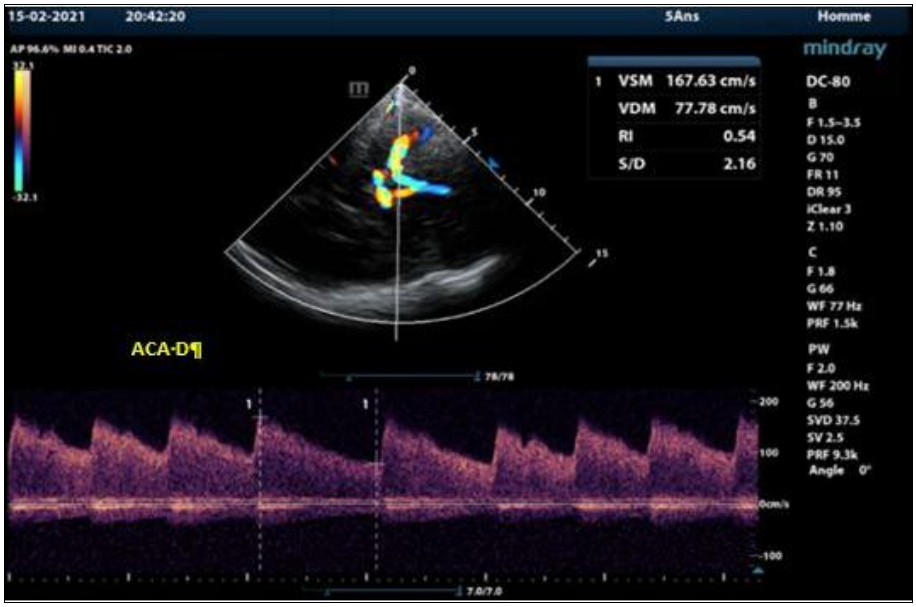

Fig N°1: velocimetric study of the right anterior cerebral artery showing an average systolic velocities equal to 167.63 cm/s.

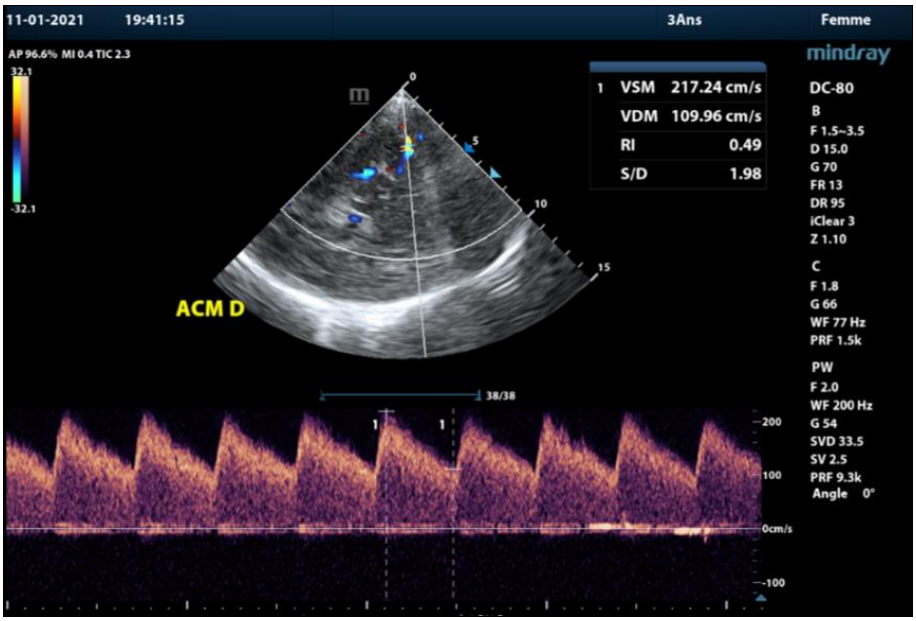

Fig N°2: Velocimetric study of the right middle cerebral artery showing an average systolic velocities equal to 217.24 cm/s.

Table 3: Distribution of patients according to the results of transcranial Doppler by age groups

Age range (Years)	Conclusion of transcranial Doppler				
	High	Low	Incomplete	Intermédiate	Total
[11-15]	12	2	2	2	18
[1-5]	36	7	3	5	51
[6-10]	38	9	--	5	57
Total	86	18	10	12	126

Conclusion

In Niger the prevalence of this disease in Niger is 25%. The most common form in the world is the homozygous form, it is a real public health problem due to its complications and one of the major complications is stroke. We therefore carried out a study on the velocimetry of the cerebral arteries in sickle cell patients with SS in order to find patients at risk of developing cerebral vasculopathy. At the end of our study, we noticed that children in the age group of 1-10 years are more likely to present high velocities, all sickle cell patients whether SS or SC can have a stroke, hence the importance of doing DTP in the follow-up of

sickle cell patients in Niger.

Conflict of interest
The authors declare that they have no conflict of interest.

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