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Original article

Pain management in children with major sickle cell syndromes (MDS) at the Albert Royer Children's National Hospital in Senegal

Prise en charge de la douleur chez des enfants porteurs de syndromes drépanocytaires majeurs (SDM) au Centre hospitalier national d'enfants Albert Royer du Sénégal

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Résumé

La drépanocytose est une pathologie fréquente au Sénégal, caractérisée par des douleurs aiguës lors des crises vaso-oclusives (CVO), qui représentent un motif majeur de consultation et d'hospitalisation. Cette étude vise à évaluer la prise en charge de la douleur chez les enfants porteurs de syndromes drépanocytaires majeurs (SDM).

Cette étude rétrospective a été réalisée chez des patients suivis pour SDM à l'unité de soins ambulatoires pour enfants et adolescents drépanocytaires (USAD) du Centre Hospitalier National d'Enfants Albert Royer (CHNEAR) de Dakar, entre le 2 janvier et le 31 mars 2023. Nous avons inclus tous les enfants de moins de 15 ans consultés aux urgences de l'USAD et ayant bénéficié d'une prise en charge de la douleur.

Nous avons analysé 100 dossiers de SDM, dont 57 étaient des filles, entraînant une sex-ratio de 0,75. La classe d'âge la plus représentée était celle des 6 à 10 ans (45 % des patients). La douleur était le mode de révélation le plus fréquent (84 % des enfants). En 2023, 11 enfants avaient plus de 3 hospitalisations pour douleur, principalement localisée au niveau des membres (70 %). L'évaluation de la douleur a été effectuée chez 37 % des enfants lors de la consultation. L'association d'antalgiques de palier I (paracétamol) et de palier II (Tramadol) a été utilisée chez 39 enfants. Le délai d'administration de la première dose était estimé à 15 minutes chez 67% des patients, tandis que l'évaluation post-antalgique n'était réalisée qu'après 5 heures pour 41 enfants.

Mots-clés : douleur, enfant, drépanocytose, Sénégal.

Abstract

Sickle cell disease is a common pathology in Senegal, characterized by acute pain during vaso-oclusive crises (VOCs), which represent a major reason for consultation and hospitalization. This study aims to evaluate pain management in children with major sickle cell syndromes (MDS).

This retrospective study was carried out in patients followed for SDM at the outpatient care unit for

children and adolescents with sickle cell disease (USAD) of the Centre Hospitalier National d'Enfants Albert Royer (CHNEAR) in Dakar, between January 2 and March 31, 2023. We included all children under 15 years of age who were consulted in the USAD emergency department and who had received pain management.

We analyzed 100 SDM records, 57 of which were girls, resulting in a sex ratio of 0.75. The most represented age group was 6 to 10 years old (45% of patients). Pain was the most common mode of revelation (84% of children). In 2023, 11 children had more than 3 hospitalizations for pain, mainly localized to the limbs (70%). Pain assessment was performed in 37% of children at the consultation. The combination of level I (paracetamol) and level II (Tramadol) analgesics was used in 39 children. The time to administration of the first dose was estimated at 15 minutes in 67% of patients, while the post-analgesic evaluation was only performed after 5 hours for 41 children.

Keywords: pain, child, sickle cell disease, Senegal.

Introduction

Sickle cell disease is a genetic disease, defined by the presence of an abnormal hemoglobin called hemoglobin S (HbS). According to the World Health Organization (WHO), nearly 5% of the world's population is a carrier of a gene responsible for an abnormality in hemoglobin [1]. Indeed, sickle cell anemia is the most common hemoglobinopathy in the world. According to the WHO, more than 330,000 children are born with this disease each year [2].

The majority of people with this disease live in sub-Saharan Africa, with prevalence rates ranging from 10 to 40% [3]. It poses a real public health problem in developing countries. In Senegal, one in ten people, regardless of ethnicity, geographical origin or social class, is thought to be a carrier of the sickle cell gene, with 0.5% of annual births of major forms [9,10]. Transmission is autosomal recessive. Indeed, people who have inherited the gene from only one of the parents do not classically show clinical signs; they are carriers of the sickle cell trait (SMA) and are asymptomatic. Those who have inherited the gene from both parents have the homozygous SS form, which is the most common of the major sickle cell syndromes and requires regular follow-up due to clinical manifestations. The most common of these manifestations is the pain of the vaso-oclusive crisis (VOC) which is the main symptom of the disease, with bone and/or abdominal localizations. These are painful crises that can occur spontaneously, or triggered by favourable circumstances: cold, heat, sudden changes in temperature (e.g. cold water bath), fever, dehydration (vomiting, diarrhoea), altitude, lack of oxygen, intense and sustained physical exertion, clothing that is too tight, obstruction of the airways, systemic corticosteroid use [4,5].

Pain management in patients should take into account the duration of pain progression and intensity, using an appropriate scale [6]. Thus, pain can be mild, moderate or severe, transient or prolonged over several days, with single or multiple locations (spine, limbs, chest, abdomen) [6]. However, from a biological point of view, no examination is necessary. There are no specific markers to identify CVO or assess its severity [6]. In children, the pain of the CVO is the most frequent reason for consultation in the emergency department [7]. Management requires an assessment of its intensity using validated self-assessment or heteroassessment scales, a prescription of appropriate analgesics and monitoring of the effectiveness and tolerance of analgesics. Indeed, the relief of this pain is the main objective of the practitioner and the only expectation of the patient. However, it is important to remember the difficulty of assessing and relieving pain in some children, in certain contexts. It is in this context that our work is carried out, the objective of which was to evaluate the management of pain in children with SDM and, more specifically, to describe the socio-demographic characteristics of patients, to specify the main locations of pain, to evaluate the use of pain scales, as well as the time taken to administer the 1st analgesic.

Methodology

We conducted a retrospective, descriptive and cross-sectional study at the Albert Royer Children's National Hospital (CHNEAR) in Senegal over a period from January 2 to March 31, 2023, i.e. a period of 3 months. Our work focused on children with major sickle cell syndrome followed at the outpatient care unit for children and adolescents with sickle cell disease (USAD) in Senegal.

We included all children under 15 years of age, followed for major sickle cell syndrome confirmed on hemoglobin electrophoresis, received in emergency consultation at the USAD and received pain management.

Data were collected on a survey sheet, from the consultation register and the patient's medical follow-up records.

The variables studied were:

- socio-demographic parameters: age, sex;
- the characteristics of sickle cell anemia: type, circumstances of discovery, onset of follow-up, complications, etc.)
- Pain assessment and management (scales, analgesics, monitoring).

Results

During our study period, we included 100 children followed for SDM who had consulted for pain and were being treated at the USAD.

- **Socio-demographic parameters**

Age

The most represented age group was [6 – 10 years], with 45% of patients. (Table I)

Sex

Our population was made up of 57 girls, a sex ratio of 0.75.

- **Characteristics of sickle cell disease**

Type of major sickle cell syndrome

Homozygous sickle cell disease was the most common in our cohort with 93% of children (Table II).

Mode of Sickle Cell Disease Disclosure

Pain was the most common mode of revelation (84% of children) (Figure 2).

Number of hospitalizations per year for pain

During the year 2023, 11 children in our cohort had more than 3 hospitalizations for pain (Figure 2).

Location of pain during hospitalization

The localization of limb pain was found in 70 children (Figure 3).

- **Pain Management**

Pain assessment by a paediatric scale

Pain assessment was performed in 37% of children during the consultation (Table III).

Analgesics used

The combination of level I analgesics (paracetamol) and level II analgesics (Tramadol) was used in 39 children (Figure 4).

Time to administration of the first analgesic dose

During our study period, 67 children received analgesics in less than 15 minutes (Figure 5).

Time to evaluation after first dose of analgesic

Assessment of pain after the first dose of analgesic was performed only after 5 hours in 41 children (Figure 6).

Table I: Patient Compensation by Age Group

Age in year	Workforce (N)	Percentage (%)
0 - 5	14	14
6 – 10	45	45
11 – 15	41	41
TOTAL	100	100

Table II: Type of Major Sickle Cell Syndrome

Type of Sickle Cell Syndrome Major	Workforce (N)	Percentage (%)
S Beta thalassemia	1	1
SC	6	6
SS	93	93
TOTAL	100	100

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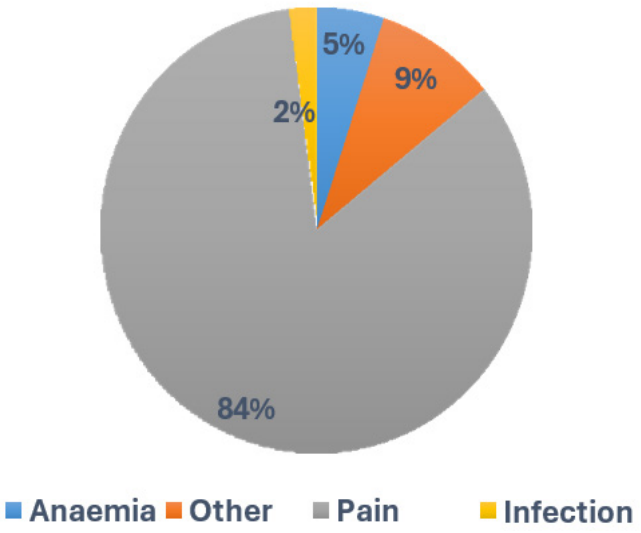

Figure 1: How sickle cell disease is revealed

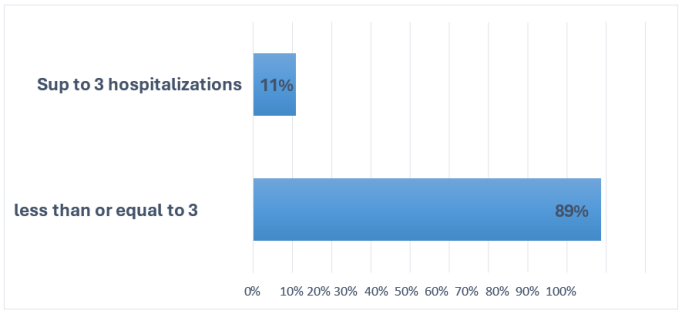

Figure 2: Frequency of hospitalization for pain per year

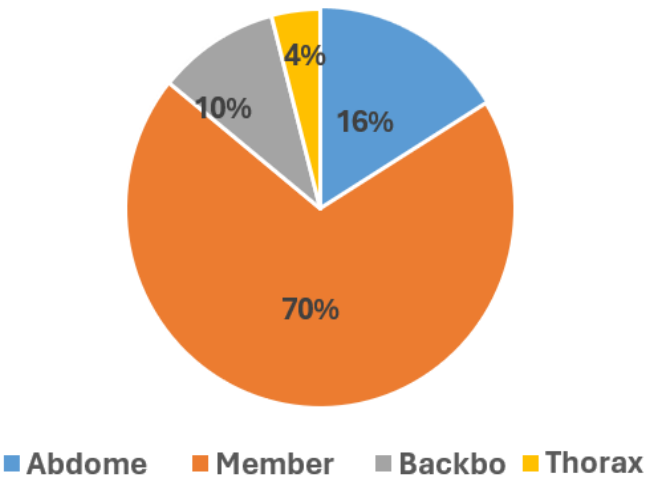

Figure 3: Location of pain

Table III: Assessment of pain using a paediatric scale

Pain Assessment/ Pediatric Scale	Workforce (N)	Percentage (%)
No	63	63
Yes	37	37
TOTAL	100	100

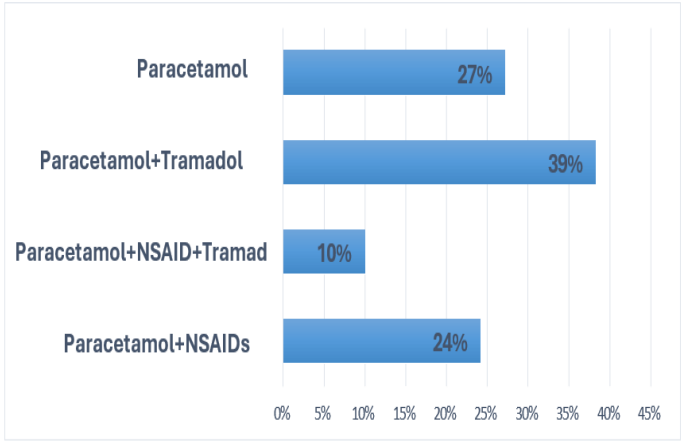

Figure 4: Analgesics used

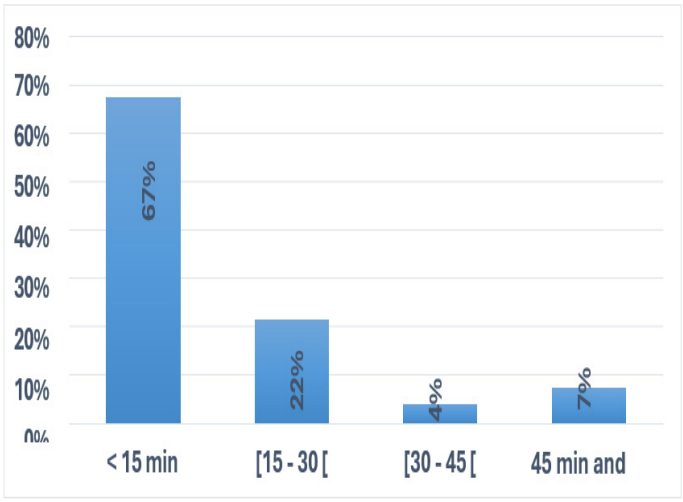

Figure 5: Time to first dose of analgesic

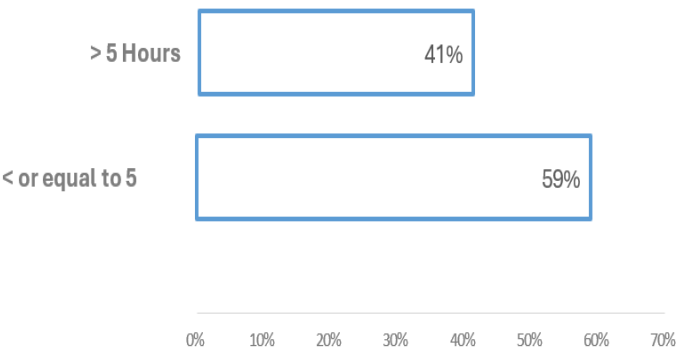

Figure 6: Time to evaluation of the first dose of analgesic

Discussion

At the end of our study, which took place at the USAD over a period of 03 months, we registered 100 patients, with a slight predominance of women (57 girls for 43 boys, i.e. a sex ratio of 0.75). Our results were comparable to those of Nacoulma et al. Females accounted for more than half of the cases (59.2%) in her study [8]. On the other hand, other authors such as Thiam et al. found a predominance of men, including 20 girls and 26 boys [9]. All these results could be explained by the autosomal recessive mode of inheritance of sickle cell disease. The age group [6 years – 10 years] was the most common with 45% of patients, followed by children aged [11 years – 15 years] with 41 patients. In 2017, Dème/ly et al., in their work on children admitted to emergency situations, found a predominance of the 5-10 age group [10]. The predominance of this age group in our study is partly due to the lack of systematic newborn screening for sickle cell disease in Senegal. As a result, the signs of the disease are expressed at the end of early childhood, corresponding to the stabilization of the fetal hemoglobin level, giving way to the beginning of signs and complications, in particular vaso-occlusive crises [11]. In addition, the Senegal haplotype is characterized by a better tolerance of the disease which is expressed later and in a more attenuated way, compared to the other haplotypes.

The most common type of MDS was homozygous SS, which was found in 93% of patients.

Thalassemia S-Beta forms were less common (1%).

This could be explained by the fact that the homozygous SS form is the most common in Senegal.

Pain was the main reason for discovering the disease (84% of patients). Similar results were observed in the study carried out by Thiam et al in 2017 in Ziguinchor [9].

In our study, 89% of patients were hospitalized one to three times a year for pain, with localization in the limbs in 70% of patients. Cissouma et al found 41.7% in the limbs, 29.1% in the abdomen and 4.2% in the chest [12].

In 63% of patients, the result of pain assessment by a pediatric scale was not mentioned in the nursing record. This means no assessment or failure to report the results of the pain assessment. This constitutes a lack of quality of care.

All patients hospitalized at USAD for pain during our study period had received analgesic treatment. The most frequently used treatment regimen was paracetamol + Tramadol, with 39% of patients. The other schemes were:

- Paracetamol alone: 27%;
- Paracetamol + NSAIDs: 24%;
- paracetamol + NSAIDs + Tramadol: 10%.

Indeed, all these molecules are level I and II analgesics. Our results are comparable to those of Gbadoé et al. who had used the same level I and II analgesics with good results [13]. Moussavou et al had recommended the wide use of level III analgesics [14]. In our patients, the lack of use of level III analgesics (morphine) was due to the unavailability of the molecule.

It should also be noted that sometimes the fear of the side effects of morphine and the reluctance of parents are limits to its use in hyper-algic CVOs in children. Indeed, in this type of very intense pain, morphine is the analgesic recommended as a first-line analgesic in a hospital setting, under supervision, especially since other lower-level analgesics are often used at home, without success. Classically in the management of bone CVO, the combination of level I and II analgesics often gives good results, in addition to intravenous hydration.

However, for patients who had abdominal pain, an antispasmodic was used in the majority of cases. The time taken to administer the first dose of analgesic was on average less than 15 minutes in 67% of patients. Regarding the time taken to evaluate the effectiveness of the treatment after the first dose of analgesic, it was estimated on average less than or equal to 5 hours in 59% of patients and more than 5 hours for 41% of patients. Normally, for optimal pain management, the evaluation should be done between 15 and 30 minutes after the first dose of analgesic, depending on the type of analgesic and the mode of administration.

In total, all of our patients had an improvement in pain after analgesic treatment.

Conclusion

The management of pain for CVO in children with sickle cell disease requires above all an organization with a well-defined care protocol. The rapid identification of these patients and the evaluation of pain using appropriate scales are essential steps in this management, and the appropriate molecules and devices must be immediately accessible to the healthcare teams. Morphine remains the main treatment for patients in cases of CVO and should therefore be widely and rapidly offered, whether orally (allowing for quick and easy administration) or intravenously. However, in our regions, the use is limited by the unavailability of the molecule, the fear of side effects but also the reluctance of parents in hyperalgic CVOs in children.

Health policies must develop programs that simplify access to analgesic molecules, particularly morphine, for rapid relief of severe pain in chronic pain pathologies such as sickle cell anemia.

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Conflict of interest : None

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