International Journal of
Health, Medicine and Nursing Practice
(IJHMNP)

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CARI Journals

International Journal of Health, Medicine and Nursing Practice
ISSN 2710-1150 (Online)
Vol.8, Issue No.2, pp 1 - 9, 2025
www.carijournals.org

Frequency and Clinical Presentation of Osteoarticular Complications of Sickle Cell Disease in Mbujimayi, DR Congo.

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Accepted: 11st Apr, 2025, Received in Revised Form: 15th May, 2025, Published: 11th June, 2025

Abstract

Purpose: Sickle cell disease is the most common genetic disorder in the world. It is responsible for serious complications, including those affecting the locomotor system. This study aimed to determine the frequency of osteoarticular complications in sickle cell patients and describe their clinical presentation in Mbujimayi.

Methods: this was a cross-sectional, descriptive and retrospective study, carried out in the surgery department of the General Hospital Bonzola, in the city of Mbujimayi, in the Democratic Republic of Congo. We collected the records of 298 sickle cell patients, among whom 67 presented osteoarticular complications.

Results: the frequency of osteoarticular complications was 22.5% in sickle cell patients in Mbujimayi. The majority of our patients were male (61.2%; sex ratio = 1.2) and their mean age was 17.0 ± 10.0 years. Pain was the most common reason for consultation (37.3%). Aseptic osteonecrosis of the femoral head (40.3%) and osteomyelitis (28.3%) were the most common complications. Unloading the limb by plaster cast immobilization (76.1%), associated with analgesics and/or antibiotic therapy (53.7%) were the most applied therapeutic methods in this study.

Unique Contribution to Theory, Practice and Policy: this study shows that osteoarticular complications, represented by aseptic osteonecrosis of the femoral head and osteomyelitis, are very frequent in sickle cell patients in the city of Mbujimayi and their management remains a major daily challenge. These results are essential for the development of prevention and monitoring policies for these patients in our context.

Keywords: *Sickle Cell Disease, Osteoarticular Complications, Frequency, Clinical Presentation, Mbujimayi.*

I. Introduction

Sickle cell disease is a hereditary hemoglobinopathy with autosomal recessive transmission [1]. It results from the mutation on chromosome 11 of the sixth codon of the ß-globin chain of hemoglobin (GAG ⇾ GTG), resulting in the substitution of a glutamic acid by a valine (GLU ⇾ VAL) on the protein. The hemoglobin S thus formed is capable of polymerizing in certain circumstances such as hypoxia, causing the sickling of red blood cells, hence the term sickle-cell disease [2].

Sickle cell disease is the most common genetic disease in the world [3]. Despite its reported ubiquity character, it particularly affects sub-Saharan Africa where it is prevalent, reaching up to 25% in some regions [4].

Sickle cell disease is a real public health problem in the Democratic Republic of Congo (DRC), our country, where the World Health Organization (WHO) estimates that the rate of heterozygous subjects (AS) is 25 to 30% and that the annual incidence of the homozygous form (SS) is around 15 per 1000 births. Each year, approximately 50,000 newborns are born with the homozygous form [4,5].

Homozygous subjects for the mutation, and some compound heterozygous subjects, have a major sickle cell syndrome and are likely to develop severe complications of the disease, among which we count osteoarticular complications, which are mainly bone infarctions, dactylitis, aseptic osteonecrosis, arthritis and osteomyelitis [2,6,7]. The latter are very frequent and their appearance at a young age makes them very serious in a resource-limited environment like ours, where their management is a significant challenge.

This article aims to determine the frequency and clinical presentation of the most common osteoarticular complications in sickle cell patients in Mbujimayi.

II. Methods

Study design and setting: This was a cross-sectional, descriptive and retrospective study, carried out in the surgical department of the General Hospital Bonzola, located in Mbujimayi, Democratic Republic of Congo; from January 1, 2011 to July 31, 2021, a period of 10 years and 7 months.

Study population: it was represented by all sickle cell patients who consulted in the department during our study period.

Inclusion criteria: This study included sickle cell patients of all sexes and ages, admitted to our department for osteoarticular complications.

Non-inclusion criteria: Sickle cell patients admitted for other clinical manifestations were not included in the present study.

Sampling: was non-probabilistic.

Data collection: Data were collected using an anonymous data collection form from patient medical records, hospitalization records, and surgical reports. We exhaustively collected the medical records of all sickle cell patients diagnosed with osteoarticular complications.

Parameters of interest: sociodemographic characteristics of patients (sex and age), reasons for consultation, region concerned, types of osteoarticular complications of sickle cell disease and therapeutic modalities.

Data analysis and processing: Microsoft Excel 2010 and Jamovi 2, 2, 5 software were used. The usual descriptive statistics tests were calculated (frequency, mean and standard deviation).

Ethical consideration: This study received approval from the Faculty of Medicine of the Official University of Mbujimayi. Patient data were collected using an anonymous data collection form and treated confidentially.

III. Results

1. Frequency

The frequency of osteoarticular complications of sickle cell disease at General Hospital Bonzola during our study period was 67 cases out of 298, or 22.5%.

2. Socio-demographic characteristics of patients (sex and age).

The majority of our patients were male (61.2%, sex ratio = 1.6). The mean age of patients was 17.0 ± 10.0 years. The age group between 11 and 20 years was the most represented with 43.3% of cases (Table 1).

Table 1. Distribution of patients according to their socio-demographic characteristics.

Socio-demographic characteristics	Number (n = 67)	Percentage (%)
Sex		
Male	41	61.2
Female	26	38.8
Sex ratio: 1.6 Male/Female		
Age (years)		
1 – 10	17	25.4
11 – 20	29	43.3
21 – 30	12	17.9
≥ 31	9	13.4
Mean age: 17.0 ± 10.0 years	Extremes: 2 and 41 years old	

3. Reasons for consultation and region concerned

Pain was the most common reason for consultation, reported by 37.3% of our patients. The hip was the most affected region, with 61.2% of cases (Table 2).

Table 2. Distribution of patients according to reasons for consultation and the region concerned.

Variables	Number (n = 67)	Percentage (%)
Reasons for consultation		
Pains	25	37.3
Lameness when walking	17	25.4
Fever	14	20.9
Swelling	11	16.4
Region concerned		
Hip	35	52.2
Leg	16	23.9
Knee	5	7.4
Arm	3	4.5
Thigh	2	3.0
Ankle	2	3.0
Wrist	2	3.0
Foot	1	1.5
Elbow	1	1.5

4. Osteoarticular complications of sickle cell disease

Aseptic osteonecrosis of the femoral head was diagnosed in 27 of our patients, making it the most common osteoarticular complication with 40.3%; followed by osteomyelitis (all types combined) with 28.3% (Table 3).

Table 3. Distribution of patients according to the types of osteoarticular complications diagnosed.

Osteoarticular complications of sickle cell disease	Number (n = 67)	Percentage (%)
Aseptic osteonecrosis of the femoral head	27	40.3
Chronic osteomyelitis	12	17.9
Osteoarthritis	8	11.9
Acute osteomyelitis	7	10.4
Fractures of limb bones	5	7.5
Septic osteoarthritis	4	6.0
Osteitis	3	4.5
Ankylosis	1	1.5

5. Therapeutic modalities of osteoarticular complications of sickle cell disease in Mbujimayi.

Plaster immobilization was used as the therapeutic modality of choice in this study (76.1%), generally associated with analgesics and antibiotic therapy in 53.7% of patients (Figure 1).

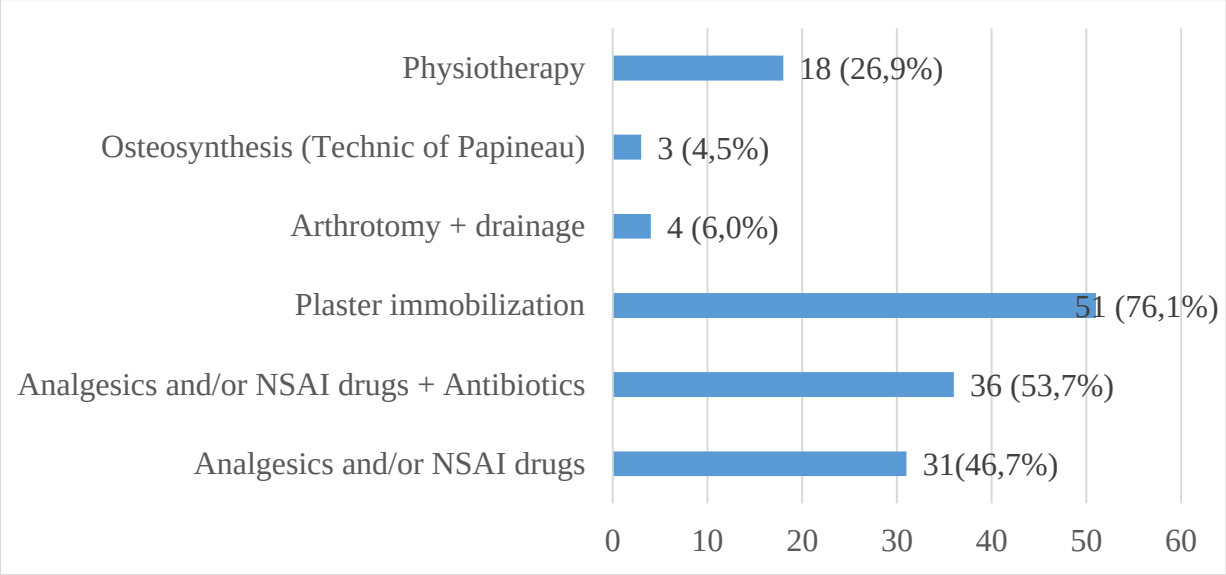

Figure 1. Therapeutic modalities for osteoarticular complications of sickle cell disease.

IV. Discussion

Frequency

The frequency of osteoarticular complications of sickle cell disease at General Hospital Bonzola during our study period was 67 cases out of 298, or 22.5%. The frequency found in this study is similar to that of Zavier Zomalhèto (25.6%) [8] and is between those reported by Diakité Mamady et al. (15%) and Diakité A. et al. (41.1%) [9, 10]. This frequency varies from one study to another. In all cases, it remains high and reflects the undoubted association that exists between sickle cell disease and the osteoarticular lesions to which it strongly exposes.

Socio-demographic characteristics

The majority of our patients were male (61.2%; sex ratio = 1.6). Although Zavier Zamalhèto and Touré noted like us a male predominance in their studies carried out respectively in Cotonou (54.5%) and in Abidjan (53.0%) [8, 11]; several other studies report practically the opposite. Indeed, several authors mention the female predominance, sometimes clear, across their series; this is notably the case Antoine De Gheldere et al. (51.7%) or Taciana Fernandes et al. (67.3%) [12, 13]. This disparity can probably be explained by the distribution of the population which is very different from one region to another.

The mean age of our patients was 17.0 ± 10.0 years. The age group between 11 and 20 years was the most affected with 43.3% of cases. We did not note significant differences in the age of patients across the literature because the mean generally swims around 15 years as reported by Touré (14.6 years) and Coulibaly et al. (17 years) [11, 4]. This result confirms the fact that osteoarticular lesions are very early in the course of sickle cell disease. In addition, it is an indicator of the life expectancy of this group of patients, only a minority of whom reach the age of 40 in Mbujimayi.

Reasons for consultation and region concerned

Pain was the most common reason for consultation, reported by 37.3% of our patients. This result is also reported by Diakité Mamady et al., all of whose patients consulted for pain [9]. Although it is the main complaint of the majority of our patients, pain remains underrepresented in our series compared to others. This observation could be explained by self-medication and the use of traditional medicine as a first-line treatment by our patients, which alleviates pain and relegates it to the background.

Osteoarticular complications of sickle cell disease

Aseptic osteonecrosis of the femoral head was diagnosed in 27 of our patients, making it the most common osteoarticular complication with 40.3%; followed by osteomyelitis (all types combined) with 28.3%. This result is comparable to that reported by Diakité Mamady, in whom 44.4% of patients presented with aseptic osteonecrosis of the femoral head, although it was not the first complication, since in this same study, osteomyelitis was diagnosed in 55.6% of patients [9]. A result which was also reported by Diakité A. et al., in whom 40.5% were affected by osteomyelitis [10]. As mentioned previously, self-medication, particularly the excessive use of antibiotics in our environment, curbs a proportion of osteomyelitis whose diagnosis is made late. Aseptic osteonecrosis of the femoral head is more frequently diagnosed in Mbujimayi because of its persistent, disabling symptoms that do not resolve using conventional methods; which ultimately pushes patients to seek medical advice. Furthermore, its high frequency explains why the hip was the most affected region (61.2%) in this study, a result that is supported by Ayed et al. [15].

Therapeutic modalities of plaster immobilization was used as the therapeutic modality of choice in this study (76.1%), generally associated with analgesics and antibiotic therapy in 53.7% of patients. Although surgery and rehabilitation physiotherapy occupy a prominent place in the management of osteoarticular complications in sickle cell patients as reported in the literature [16-19]; they were rarely used in this study, even when their indication was given, particularly in cases of aseptic osteonecrosis of the femoral head where hip arthroplasty was frequently considered.

This demonstrates the difficulty practitioners face in caring for this category of patients in our underprivileged context, where technical support and health coverage are severely lacking. On the other hand, immobilization, adequate antibiotic therapy, and analgesia adapted to each case have been of great use, as can also be read in the study published by Akakpo-Numado et al. [20]. The evaluation of therapeutic results of these different methods should be the subject of future publications in Mbujimayi.

Conclusion

Osteoarticular complications are very common among sickle cell patients in Mbujimayi. They are mainly represented by aseptic osteonecrosis of the femoral head and osteomyelitis. Their management involves usually plaster cast immobilization, combined with analgesics and antibiotic therapy appropriate to the case.

State of Knowledge on the Subject

- Sickle cell disease is the most common hereditary blood disorder worldwide and is highly prevalent in the sub-Saharan region.
- It is responsible for serious complications, both acute and chronic, including osteoarticular complications.
- To date, there is no study published in Mbujimayi, drawing up the epidemiological and clinical profile of osteo-articular complications of sickle cell disease.

Unique Contribution to Theory, Practice and Policy:

- Our study provides an epidemiological and clinical profile of osteoarticular complications of sickle cell disease in our setting.
- These results can serve as a basis for comparison with other regions to understand inter-regional variability.
- They can also serve as a basis for developing prevention and follow-up policies for these patients in our context.

Conflict of Interest Declaration

All authors declare that they have no conflict of interest.

Authors' contributions

Protocol design and writing: TKK, PCK and EMB. Data collection and compilation: PCK, GNT and HBTK. Data analysis and interpretation: TKK, DMK and HKN. Manuscript writing: TKK, SBK, AMY. Manuscript revision: MNE, PTT, DMK, HKN and MKK. All authors reviewed, read, and approved the final version of this manuscript.

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