PAIN MANAGEMENT AMONG ADULT SICKLE CELL DISEASE PATIENTS WITH VASO-OCCLUSIVE CRISES AT A UNIVERSITY TEACHING HOSPITAL

Jimoh AO¹, Ndakotsu MA², Bakare TA³, Abubakar SB⁴

Departments of Pharmacology & Therapeutics¹ and Haematology & Blood Transfusion²,
College of Health Sciences, Usmanu Danfodiyo University Sokoto.

Department of Psychiatry³ and Haematology⁴, Usmanu Danfodiyo University Teaching Hospital Sokoto.

Correspondence: Dr Abdulgafar O. Jimoh,

Departments of Pharmacology and Therapeutics, College of Health Sciences,
Usmanu Danfodiyo University Sokoto.

Email: layidotcom@yahoo.com

ABSTRACT

Background: The pain accompanying vaso-occlusive crisis (VOC) in sickle cell disease (SCD) patients is often severe, but poorly managed in many health care settings. According to the World Health Organisation report, in adequate management, absence of national control programmes, and basic facilities to manage the patients remains a challenge in most countries where sickle-cell anaemia is a major public health concern.

Objectives: This study aimed to assess the types of therapeutic intervention and pain assessment during hospitalization of sickle cell disease patients presenting with pain-related vaso-occlusive episodes (VOE).

Materials and Methods: A cross sectional retrospective review of pain management in vaso-occlusive crises was carried out. Information was obtained from 139 case files of adult sickle cell disease patients managed for VOC in the Haematology unit of the Usmanu Danfodiyo University Teaching Hospital Sokoto from June 2009 to July 2011.

Results: The mean age of respondents was 20.07±4.07 years, with females accounting for 65.5%. About 10% were married, and only 1.4% were gainfully employed while 88.5% were students. Among the patients managed for vaso-occlusive crises, majority were treated with DF118 (41.0%), followed by Non-Steroidal Anti Inflammatory Drugs (38.9%) and Paracetamol (35.3%). A few of the patients were treated with narcotic analgesic (12.2%) and non-narcotic analgesic (11.5%). About 16% of the patients were managed with intravenous fluid. HbSS was the most predominant genotype accounting for 92.3%, HbSC was 4.4%, while HbSS+F was 3.3%. Infections was the chief precipitant of the crises accounting for 66.2%, followed malaria which accounted for 33.8%.

Conclusion: There is need to optimise management of pain in vaso-occlusive crisis to conform to recommended guidelines, there was no objective tool of pain assessment, and the use of hydroxyurea in crises management was not frequently employed.

Keywords: Pain management, Vaso-Occlusive Crises, Sokoto

INTRODUCTION

Sickle Cell Disease (SCD) is the most common genetic disorder of haemoglobin in sub-Saharan Africa.^{1,2} In Africa it is estimated that about 200,000 children are born with the disease annually.³

A painful sickle cell crisis, also known as vaso-occlusive crisis is the commonest manifestation of SCD and it is characterised by recurrent episodes of acute, severe pain from tissue ischaemia as a result of vaso-occlusion.⁴ Frequent VOCs are associated with early mortality, and

can lead to chronic and intractable pain syndromes, greatly impacting sleep, mood, and social and physical functioning.⁵ Pain accounts for about 90% of hospital admission in patients with SCD,⁶ and also remains the major concern of patients and their families.⁷

Acute pain management in the sickle cell disease (SCD) individuals is often initiated by attending health care professionals including

Haematologists. Anaesthetists as pain specialists are sometimes involved in the management of painful vaso-occlusive crises and they also play major roles when Intensive Care Unit management is required.⁸

"Pain ladder" is a term coined by the World Health Organization (WHO) to describe its guideline for the use of drugs in the management of pain. It was originally applied to the management of cancer pain, but is now widely used by medical professionals for the management of all types of pain.⁹ According to this guideline, patients who present with mild to moderate pain should be treated with a non-opioid analgesic. An adjuvant drugs should be used if a specific indication exist. Adjuvant drugs include treatments for opioid side effects or other comorbid conditions and "adjuvant analgesics," which are drugs with primary indications other than pain that are analgesic in selected circumstances.⁹ Patients who fail the 1st-step regimen or who present with moderate pain should be treated with an oral opioid for moderate pain as well as with a non-opioid analgesic and an adjuvant drug.⁹ Patients who fail the 2nd-step regimen or who present with moderate to severe pain should be treated with an opioid indicated for such pain. The clinician should consider the addition of a non-opioid analgesic or an adjuvant

drug. In the treatment of persistent pain, analgesics should be given on a regular basis—"by the clock"—so that the next dose is given before the effect of the previous one wears off.⁹

Pain frequently occurs spontaneously, but may be precipitated by infection, exposure to cold, dehydration or in uncommon situations after emotional stress, exercise or alcohol, often no precipitating cause can be found.^{4,6} In sickle cell anaemia patients older than 20 years, an increased frequency of pain is associated with early death.¹⁰ How well or poorly this pain is treated has important consequences as to how patients cope with pain and life. Unfortunately, few studies have examined the treatment of painful crises among this group of patients.

This study aimed to assess the types of therapeutic intervention during hospitalization of sickle cell disease patients presenting with pain-related vaso-occlusive episodes (VOE). We also examine the common precipitating factors of vaso-occlusive crisis and the types of sickle cell genotype among the patients.

MATERIALS AND METHODS

This was a retrospective review of the clinical records of sickle cell disease patients in vaso-occlusive crises managed at the Haematology unit of the Usmanu Danfodiyo University

Teaching Hospital, Sokoto State. The patients were treated between June 2009 and July 2011 as in patient. The Usmanu Danfodiyo University Teaching Hospital, Sokoto serves as a referral centre for specialist and general hospital in the state. The Haematology unit has 2 Honorary Consultants, 3 visiting Consultants and 2 Senior Registrars. The unit operates daily emergency services and one day outpatient clinic per week. There are both inpatient and outpatient facilities in the unit.

The study involved sorting and retrieval of the case notes of the 139 patients who had vaso-occlusive crisis in the haematology unit from the record department. A proforma questionnaire was designed by the researchers and the following data were recorded: Bio-data, pattern of pain assessment, pattern of pain management, drugs used by the patients, drug used in the management of pain associated with vaso-occlusive

Table 1: Socio-demographic factors of the patients

Variables	no(%)
Age (Years)	
10-14	6(4.3)
15-19	63(45.3)
20-24	50(36.0)
25-29	18(13.0)
≥30	2(1.4)
Sex	
Male	48(34.5)
Female	91(65.5)
Occupation	
Student	123(88.5)
House wife	14(10.1)
Employed	2(1.4)
Marital status	
Single	126(90.7)
Married	13(9.3)

Figure 1. Respondents perception of severity of pain **Figure2:** Percentage distribution of genotype **Table 2: Pattern of pain management**

Mild-moderate crisis (n=117)		Moderate-severe crisis (n=22)	
Drugs used	n(%)	Drug used	n(%)
Pentazocine	5(4.2)	Pentazocine	12(54.6)
DF118	46(39.3)	DF118	11(50.0)
NSAIDs	38(32.5)	NSAIDs	16(72.7)
Paracetamol	38(32.5)	Paracetamol	11(50.0)
Tramadol	7(5.9)	Tramadol	9(40.9)
		Hydroxyurea	8(36.4)

crisis, types of sickle cell disease genotype and the precipitants of the pain crisis.

Data analysis

The data collected was entered into and analysed using the Statistical Package for Social Sciences (SPSS) version 20. The results were presented using frequency tables, percentages and charts for all categorical variables.

RESULTS

A total of 139 case files were reviewed. The age ranged between 10 and 32 years (20.07 ± 4.07 years). Forty-eight (34.5%) were males, 88.5% were students, while only 1.4% were employed and 90.7% were single (Table 1).

No tool was used for pain assessment during the course of vaso-occlusive crises pain management in these sickle cell disease patients. All the twenty-two (15.8%) patients that were managed for moderate-severe crises had intravenous fluid while on admission. Among the 22 patients that presented in moderate-severe crisis, less than half (45.5%) were not administered narcotics analgesics, 50% were administered DF118, majority (72.7%) were administered NSAIDs, 50% were administered Paracetamol, only 8 (36.4%) were treated with Hydroxyurea and less than half (40.9%) were administered Tramadol. Among patients with mild-moderate crisis, none of them was managed with intravenous

fluid, 4.2% were administered narcotics analgesic, 39.3% were administered DF118, 32.5% were treated with NSAIDs, 32.5% were treated with Paracetamol and only 5.9% were treated with Tramadol. Overall, 12.2% were treated with narcotic analgesics, 41% were treated with DF118, 38.9% were treated with NSAIDs, 35.3% were treated with Paracetamol, while 11.5% were treated with Tramadol. The HbSS genotype was most predominant accounting for 72.7%, HbSC was 5.7%, while HbSS+F was 21.6%. Infections was documented in the case files as the commonest precipitant of vaso-occlusive crises accounting for 66.2%, followed by Malaria which accounted for 33.8%.

DISCUSSION

Pain from a vaso-occlusive crisis is often undertreated because of concerns about narcotic addiction and tolerance, perceived drug-seeking behaviour, excessive sedation, respiratory depression and lack of specific findings on the physical examination.¹¹⁻¹³

This hospital study observed that patients who presented with mild-moderate crisis were not managed with intravenous fluid, few of them were administered non-opioid analgesic, DF118, NSAIDs and Tramadol. Use of opioid analgesic was not common among patients with mild-moderate crisis, while Hydroxyl urea was not used.

Also majority of patients who presented with moderate-severe pain crisis were all managed with intravenous fluid in addition to the use of opioid analgesic. This pattern of pain management fairly agrees with the three-step "analgesic ladder" recommended by the World Health Organization (WHO) for the treatment of cancer-related pain.¹⁴ The choice of analgesic and the dosage used should be individualized based on severity of pain. According to WHO, patients with mild pain can often be treated at home with oral fluids and non-opioid analgesics.^{13,15} Patients can also be started on Acetaminophen with or without Codeine or Oxycodone (Roxicodone), depending on pain severity. Non Steroidal Anti-Inflammatory Drugs can be used unless they are specifically contraindicated because of peptic ulcer disease, renal disease or hepatic dysfunction.¹¹

Narcotic analgesics can be used in patients with moderate to severe pain. The dosage of the selected narcotic should be titrated to achieve effective pain control.^{11,13,15} Because of the dose-limiting side effects of weak opioids (Codeine and Oxycodone), which include sedation, nausea and vomiting, these drugs are best used to manage moderate pain. Pain that is sufficiently severe to require an emergency department visit or hospitalization should be

treated with stronger opioids^{16,11,13,15,17}

Interestingly, various other medications like Multivitamins, Antihistamine, Antiemetic and Sedatives were used among the patients. This finding agrees with the WHO recommendation that adjuvant nonopioid agents such as Antihistamines and Antiemetics can be helpful for preventing or relieving opioid-related side effects.^{13,15}

The use of other medications like Antidepressant, Anxiolytics is also in keeping with recommendation that adjuvant analgesics such as Tricyclic Antidepressants should be considered in patients with a chronic pain syndrome resulting from recurrent acute pain crises.¹⁸

It has also been reported that nonpharmacologic techniques can be tried. These measures include physical therapy, rest, heat application, Transcutaneous Electrical Nerve Stimulation (TENS), self-hypnosis and diversional techniques.^{11,13,15,20}

Hydroxyurea was not commonly used among the patients in this study. A study reported that Hydroxyurea can be used in SCD patient to achieve great benefit in preventing painful episodes.¹⁹ It is worthy of note that the use of Hydroxyl urea comes with caution because of the risk of neutropenia.

CONCLUSION

We found that the current pain management practices of patients in vaso-occlusive crises in the haematology unit of the hospital fairly comply with the WHO guideline. However, there is a need for strict adherence to use of objective tool for pain assessment in the course of management. There is a need for current WHO guideline recommendations for pain management to be readily available to all physicians involved in pain management, as this may improve the quality of service.

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