

Full Length Research Paper

A study of the management of sickle-cell patients in the University of Nigeria Teaching Hospital, Enugu, Nigeria

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Sickle cell anemia is a progressive hemoglobinopathy producing chronic hemolytic anemia, microvascular thrombosis, ischemic pain, tissue infarction, decreased quality of life, shortened life expectancy. The study evaluated health profiles of sickle-cell anaemia (SCA) patients. This simple random cross-sectional survey sampled 68 subjects undertaking treatment at the Sickle-Cell Clinic, University of Nigeria Teaching Hospital, Ituku-Ozalla between January to December, 2010; using an adapted and modified questionnaire which also used Visual Analogue scale to assess the pain levels. After which, data collected were analysed descriptively. 30(44%) and 38(56%) subjects were males and females respectively. 13(19.0%) SCD patients within <1-5years were most frequently treated and 2(2.9%) of 31-35 & >45years-the least. Prevalence of pain occurred among the entire 68 SCA patients. Pain provocative factors include Stress 53 (77.9%), Sitting 4(5.9%), and cold 2 (2.9%). 62 (91.2%) respondents reported direct proportional relationship between pain and Activities of Daily Living; with complications in all patients. Experts' management involved 68(100%) haematologist and 9 (13.2%) physiotherapists. Patients' perception of pain relief recorded 61(89.7%) temporal; anger and depression respectively recorded 1 (1.5%) post-treatment; and non in 5 (7.4%). Conclusively, pain perception was an important psychologic and therapeutics factor for the patients wellbeing post-treatment. However, SCA patients' pain management required qualitative and clinical evidence-based multiagency healthcare services approach especially physiotherapy.

Keywords: Profiles, pain perceptions, management, sickle-cell patients, multiagency healthcare approach, Southeast and Nigeria.

ABBREVIATIONS

University of Nigeria Teaching Hospital, Ituku-Ozalla, (UNTH), Enugu, Sickle Cell Disease (SCD), Sickle-Cell Anaemia (SCA) and Health Related Quality of Life (HRQOL).

INTRODUCTION

Sickle cell anemia is a progressive hemoglobinopathy producing chronic hemolytic anemia, microvascular thrombosis, ischemic pain, tissue infarction, decreased

quality of life, and ultimately shortened life expectancy (Miller, 2013; Vichinsky et al., 2000; Gladwin et al., 2004; Shapiro, 1989; DeBaun et al., 2012; Chiang et al., 2005). It is a disease found to shorten life expectancy and cripple the social integration of individuals. It is an inherited or genetic blood disorder affecting the hemoglobin. It has giving a lot of concern to the world at large and seen as almost solely a chemotherapeutic case; that threatens the social integration and quality of life of affected individuals. SCA causes the red blood cells to lose their flexibility and disc shape, becoming stiff and curved like a sickle, thereby leading to risks of various complications. Hence, patients become vulnerable to infections, jaundice, hand-foot syndrome and entire body pain episodes; and present symptoms

and various complications (avascular necrosis, acute chest syndrome, stroke, priapism and osteomyelitis (Vichinsky et al., 2000; Gladwin et al., 2004; Shapiro, 1989; DeBaun et al., 2012).

However, since the red blood cells have an unusual shape, they can easily clump together and form small blood clots in small blood vessels. These micro thrombi can cause significant pain (Gladwin et al., 2004; Wrontniak et al., 2012). Invariably, since pain is one of the complications, it impinges on the functions of SCA patients during crisis secondary to avascular necrosis (Vichinsky et al., 2000; Gladwin et al., 2004; Shapiro, 1989; DeBaun et al., 2012).

The pain experienced during a sickle cell crisis is the product of the Sickled and less malleable red blood cells adhering to the vascular endothelium, plugging arterioles and capillaries and ultimately leading to occlusion and infarction. This pain caused can be acute, chronic or a mixture of the two. The acute pain of tissue infarction in skeletal or soft tissue tends to be sudden, unpredictable, onset and intense. However, after the resolution of sickle cell crisis, it usually stops (Okpala, 2002). Chronic pain in sickle cell disease is not simply a continuation of the pain of vaso-occlusion, the most common cause of Hospitalization of patients with sickle cell anaemia (Elander, 2013). It is usually secondary to avascular necrosis of the bone at various joints namely: the hip, shoulders, and ankle joints in decreasing order of frequency. Also, avascular necrosis commonly develops in the spine, causing chronic back pain and displaying the well known 'fish mouth' appearance on the X-rays; as the knees are seldom involved. The SCA individuals are not immune to other painful acute or chronic disorders unrelated to the haemoglobinopathy. Similarly, chronic joint pain in sickle cell patients can be caused by rheumatoid arthritis, osteoarthritis, or other degenerative joint diseases (Vichinsky et al., 2000; Gladwin et al., 2004; Shapiro, 1989; DeBaun et al., 2012; Okpala, 2002).

Perception of pain refers to the feeling the patients have at the level of their pain after treatment (Wrontniak et al., 2012; Wright et al., 2010; Shapiro et al., (2013); McClish et al., 2005). The health quality of life of the sickle cell anemia patients is a major concern and a measure of the level of pain perception, psychologic factor and the outcome of the management of crises. According to Chiang et al., (2005); McClish et al., (2005); Yale et al., (2000), the resilient psychological functioning in pediatric sickle cell disease (SCD) is now considered the norm. There is also general agreement that health-related quality of life (HRQOL) is impaired among these youths. Sickle cell pain, a common manifestation that is recurrent, acute, and unpredictable, may be the most important disease

complication associated with decrements in physical and psychosocial domains of health-related quality of life; thereby, making improved health-quality of life an important objective of medical care.

The sickle cell patients are seen as very unstable and restricted, even though most-times managed with chemotherapy. The recent developments have deemed it fit for other means like physiotherapy other than chemotherapy to be employed in the management of sickle cell patients, especially as regards joint pain and pain crisis. The goal of health care providers remains to enhance treatment outcome and restore comfort and well being to the lives of patients. However, the patients' quality of stability of life is a function of pain management, life and pain coping ability, and level of perceptions on pain management.

Nevertheless, how can this pain management be improved one may ask? Until recently management of sickle pain was the sole province of hematologists. But, a recent National Confidential Enquiry into patient outcome and death reports with other highlighted problems emanating from the management of pain with opiod analgesia for Sickle cell anemia. Consequently, it was suggested that a multi-agency approach similar to that used in palliative care be applied (Wrontniak et al., 2012; Wright et al., 2010; Porter et al., 2012). Therefore, it is imperative to determine the health profile and management of pain among sickle cell patients in University of Nigeria Teaching Hospital, Southeast Nigeria. However, it is believed that the outcome of this study would be beneficial in: improving the referral system functionally as regards sickle cell patients; understanding how sickle patients perceive of pain relief and how it affects their lives and thus show more ways through which the health-related quality of their lives will be improved.

MATERIALS AND METHODS

Participants and study site

68 regular Sickle Cell Anaemia patients (within the age ranges of <1->45years old, of both genders-table1); that are undergoing treatment at the Clinic from January-december, 2010) at the Sickle Cell Clinic of the premier University of Nigeria, Teaching Hospital, Ituku-Ozalla, Enugu established in the 1960s. The clinic serves and covers the southern and middle belt regions of the country. As an interdisciplinary teaching hospital, it offers teaching, training, and research opportunities for students, house-officers, health personnel, residents, and practicing opportunities for consultants; and clinical trials for local or international Commercial Research Organizations. This interdisciplinary clinic alone is staff

Table 1. Demographic data (Age & Sex-Frequency and Percentages)

Age range	Frequency	Percentages
<1-5years	13	19.0
6-10years	7	10.2
11-15years	9	13.2
16-20years	10	14.7
21-25years	6	8.8
26-30years	3	3.4
31-35years	2	2.9
36-40years,	7	10.2
41-45years	9	13.2
>above 45years	2	2.9
Total	68	100.0
SEXES	Males	30
	Females	38
		44.1
		55.9

Table 2. Respondents' rate of pain perception, perceptions differences and provocative factors

ITEMS	FREQUENCY	PERCENTAGES
A. Rate of pain perception	68	100.0
B. Different Pain Perceptions after treatments among respondents		
i. No Idea	5	7.4
ii. Temporal	61	89.7
iii. Angry	1	1.5
iv. Depressed	1	1.5
Total	68	100.0
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D. Provocative Factors of pain among the respondents		
Valid don't know	9	13.2
Stress	53	77.9
Sitting	4	5.9
Cold	2	2.9
Total	68	100
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E. Relationship between Pain and Activities of Daily Living (ADL) among the respondents		
i. Pain negatively affected ADL	62	91.2
ii. Pain did not affect ADL	6	8.8

by 20-25 professors and 20 consultants on full time appointment.

Sampling technique

The cross-sectional survey study implored a purposive convenience sampling method. The McGill pain properties Questionnaire (a version of Melzack's) which was adapted, modified and validated consist of 33 close-ended and open-ended questions (made up of three sections; section 1 is of demographic data, section 2 is the assessment of pain while section 3 is on the management and pattern of referral and perception of pain relief after treatment). This structured questionnaires evaluated the level of pain, treatments given using the Visual Analogue scale 1-10 (VAS) and the patients' pain-relief perception level post-treatment. However, only patients diagnosed with sickle cell anemia, presenting with pain and are undergoing treatment at the time of the study were considered for

participation. Those excluded were: patients with pain not related to sickle cell crisis, patients with visual impairment and have previously been explained on the purpose of the study. Then 8 research assistants who were medical students were trained on the instrument based on the concept of SCA, and on how to interpret SCA terminologies and concepts to the respondents. After certifying the excellent knowledge on the interpretation technique, SCA terminologies and concepts, they were assigned with the questionnaires after the purpose of the study was explained to them. Only the patients that voluntarily consented to participation were finally interviewed for the study. The approval for this study was obtained from the Department of Hematology and Human Research Ethical Committee, University of Nigeria Teaching Hospital Ituku-Ozalla, Enugu Nigeria.

Data analysis

The questionnaires were completed within three to four

Table 3. Complications associated with Sickle Cell Disease

Complications	Frequency	Percentage
Jaundice	10	10.2
Bone stress fracture	24	24.4
Swelling of the body (body abdomen & joints)	23	23.4
Dislocation	7	7.1
Avascular Necrosis	13	13.2
Musculoskeletal Disease (OA)	21	21.4
Total	98	100.0

Table 4. Experts involved in the management of pain among the SCD patients

Items	Frequency	Percentage
Phy	57	83.8
Phy & PT	9	13.3
Phy & O.S	2	2.9
Total	68	100.0

Phy = Physicians, PT = Physiotherapists, O.S = Orthopaedic Surgeons

weeks after the distribution. Data collected from the study was to evaluate the: level of pain, provocative and palliative factors, and functional performance, etc.

Descriptive analysis was done using the variables of interests using SPSS software version 16, Chicago IL, USA. The values were represented as frequencies and percentages.

RESULTS

Table 1 showed 68 respondents age ranges:13(19.1%)<1-5years, 7(10.2%)6-10years, 9(13.23%)11-15years, 10(14.7%)16-20years, 6(8.8%) 21-25years, 3(3.4%)26-30years, 2(2.9%)31-35years, 7(10.2%)36-40years, 9(13.2%)41-45years and 3(4.4%) >45years old. The highest frequency of SCA crises occurred at <1-5years-13(19.1%) 10(14.7%) 26-30years, and the least at 2(2.9%) 31-35years; genders: 38(55.9%) females and 30 (44.1%) males.

Table 2 showed the entire 68 (100%) SCA patients experienced pain during crises. On the post-treatment perception, 61(90%) respondents' reported temporal, 2(2%) patients were angry and depressed respectively; and 5(7.4%) respondents reported no perception. Pain Provocative factors among the patients were recorded as: stress 53 (77.9%), sitting 4(5.9%), and cold 2 (2.9%); and 9 (13.2%) patients nil. Table 2 also showed relationship between pain and funtional performance of SCA patients. The increased levels of pain negatively influenced the daily functional performances of 62(91.2%), while 6(8.8%) claimed non-interference on activities.

Table 3 showed complications of SCA patients as: Jaundice 10(10.2%), abdominal, joints and general body swelling 23(23.4%), dislocations 7(7.1%), stress fracture

24(24.4%), avascular necrosis 13(13.2%), musculoskeletal diseases like osteoarthritis 21(21.4%).

Finally, table 4 recorded the experts that managed SCA patients as: 57(83.8%) Haematologists, 9(13.3%) jointly by Physiotherapists and Orthopaedic Surgeons; haematologists and Orthopaedic Surgeons jointly consulted with 2(2.9%) cases.

DISCUSSION

This study reported most affected age range as <1 to 5years; but George et.al (2001) reported highest incidence age as <1-10years in UPTH (South-south Nigeria), while, Williams (2013) reported 5months to 8years of age in USA. Miller (2013) reported of 2months-5years old patients presenting SCA. This study also reported more females 38(55.9%) than males 30 (44.1%), while George and Opara (2001) reported a male/female ratio of 1.2:1 in UPTH. On pain incidence, this study reported it in all the respondents. This was confirmed by the reports of Okpala (2002); Vinchinsky et al (2000); Gladwin et al (2004); Shapiro (1989); and DeBaun et.al (2012), and specifically, the report of George et.al (2001) on Vaso-occlusive crises bone pains among 90% of SCA patients in UPTH, Nigeria. In another development, Vichinsky et.al (2000); Gladwin et.al (2004); Shapiro(1989); DeBaun et.al (2012); Okpala (2002); McClish et.al (2005); George et.al (2001) that most pain is usually secondary to avascular necrosis of the bone at various joints. The occurrence of pain, as the most prominent factor among the entire respondents of this study and other works caused SCA patients hospitalised. This finding also agrees with the study of Platt et al (1991) on the history of sickle cell disease which showed that about 5% of patients in the entire hospital as recorded

The level of pain can be assessed using visual analogue scale or pain questionnaires such as McGill pain questionnaire.

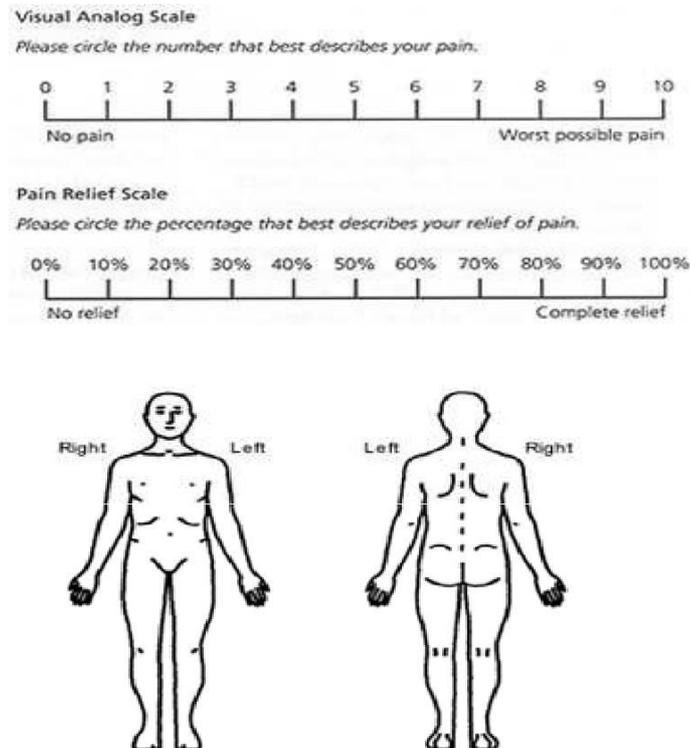


Figure1. Visual Analog and Pain Relief Scales

were devoted to pain control. Another aspect of the study result is the respondents' perceptions post-treatment, which differed among respondents ranging from temporal and permanent; anger and depressions effects with the relief relapsing after 3-4months. These results agreed with Porter et.al (2012) and Elander (2013) findings about the subjects different descriptions perceptions post treatment. These different pain descriptions existed because of different personalities and age ranges.

This study evaluation on pain provocative factors among the respondents indicted stress to cause the highest frequency of pain among SCA followed by sitting, cold and dehydration and infections. This result agrees with Shapiro (2013); Ballis et al (1996); Serjeant et al (1994) who reported on dehydration, infection and cold weather as the precipitating factors to vasoocclusive crisis. The evaluation of interference of pain with activities of daily living recorded a negative high frequency of interference with their activities of daily living. Miller (2013); Vichinsky et al (2000); Gladwin et al (2004); Shapiro (1989); DeBaun et al (2012);Chiang et al (2005) partly agreed with this finding as he earlier reported that

SCD disease shortens life expectancy and cripple the social integration of patients.

The complications identified with the SCA patients involved in this study include: Swelling at abdomen, joints, and most parts of the body, dislocations, stress fracture, avascular necrosis, and musculoskeletal diseases like osteoarthritis. This confirmed Okpala (2002); Vinchinsky et al (2000) findings Also, Barakat (2008); Vinchinsky et al (2002); McClish et al (2005); observed that these reported complications affect the resilient psychological functioning in SCD, which are considered norms and that health-related quality of life (HRQOL) are impaired in the individuals especially the young ones among them. Though, this was not reported in this study.

Finally, the professionals involved in managing the patient's crises by referral system include the Physiotherapists and Physicians. They treated the SCA cases solely and jointly just as Miller (2013) and Yale (2000) reported. Probably, the multidisciplinary approach asan option was adopted by health professionals after a solitary approach perhaps had yielded non-clinical evidence in treatment of SCA cases.

CONCLUSIONS

Pain was the patients' major challenge, making them consult with health professionals and getting hospitalized with little or no hope of living without pain. All patients willed for treatments that will ameliorate their condition.

However, the management of SCD requires Interdisciplinary evidence-based pain relief approaches. Based on the outcome of this study, it is recommended that: more studies be carried out by healthcare providers involving more time and increased sample size as to make minimal possible standard errors and ascertain the best evidence based means of sickle cell pain management, the referral systems should employ a multigency approach and counseling sessions be considered on perception of pain.

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Competing interest

The authors declare that there is no conflict of interests.

Authors' contributions

ACC as the institutional assigned supervisor to the candidate supervised the study, initiated the manuscript publication and proofread all the manuscript. ECI advised on the manuscript publication. OGC initiated and conceptualized the study as well collected the data. ESB did help in the literature reviews and periodic helped in the manuscript corrections and proofreading.

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