



Research Article

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Pattern and Severity of Vaso Occlusive Crisis in Paediatric Sickle Cell Anaemia Patients



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Abstract

Background: Sickle cell anemia (SCA) remains an inherited disease of public health significance in sub-Saharan Africa with Nigeria accounting for the significant burden of the disease globally. Acute painful crisis/event remains the hallmark of the disease with significant impact on physical and psychosocial wellbeing of both the children and the parents/caregivers despite advancement in care. The pattern and severity of acute painful event/VOC among children living with sickle cell anaemia were evaluated with a view to understand the manifestations of SCA in this group of patients.

Methods: This was a prospective cross-sectional study and involved patients who presented at the emergency room, or out-patient unit of the Paediatrics Department of the Lagos University Teaching Hospital, Idi-Araba Lagos with acute painful crisis over a nine-month period (March 2015 - November 2015).

Result: The participants mean age was 8±7 years with male to female ratio of 1.1. The distribution of pain with respect to anatomic sites were the extremities only 47(43.4%), extremities and other sites 27 (25.5%), abdomen 13(12.3%) and chest 12(11.3%). Sixty-six (62.3%) children with SCA presented with moderate pain intensity, 30% with severe pain intensity and 4.7% with mild pain intensity. Most of the children presented at the hospital within four days of pain onset.

Conclusion: The commonest site of pain among children living with homozygous sickle cell anaemia is in the extremities. Most patients presented within the first four days of onset of pain and with moderate intensity.

Keywords: : Children; Sickle cell; Pain; Severity; Location

Introduction

Sickle cell anaemia (SCA) is a homozygous form of sickle cell disease which is an inherited autosomal recessive disorder. [1,2] The disease is prevalent amongst people of Sub-Saharan Africa, Mediterranean basin, Arabian Peninsula, and India sub-continental ancestral origin. Sub-Saharan Africa however accounts for the largest burden of disease worldwide with about 200,000 of the 300,000 live births delivered with sickle cell disease yearly. [3-5] Nigeria accounts for the highest disease burden and mortality in children below the ages of five years with sickle cell anemia [6-8]. SCA is chronic hemolytic disease with an ongoing risk of acute manifestations and multi-organ dysfunctions [1,9,10].

Acute Painful episodes or vaso-occlusive crisis [VOC] remain the hallmark of SCA worldwide [11,12]. It starts at infancy and spans throughout the patient's lifetime. Its onset can be acute, subacute, chronic, or episodic and its predisposing factors include infections, extremes of weather, stress, which could be emotional, psychological, or physical. [1,9,13]. The pain itself could be somatic, visceral, neuropathic, or even iatrogenic with devastating effects on the physical, psycho-social, financial, and quality of life of the patients, care givers and the entire society due to recurrent hospitalization [11,14]. The mainstay in the management of pain in SCA includes hydration and analgesics with other pain controlling measures; this is in addition to plausible treatment

for the predisposing conditions which include administration of appropriate antibiotics to infection, treatment of malaria, red cell transfusion, and the provision of adequate psychosocial support [13,15-17]. Similarly, there are preventive measures that could be taken against pain and other vaso-occlusive crises, these include adequate oral fluid intake, avoidance of extremes of weather and stressful events, the use of antimalarial and pneumococcal prophylaxis, vaccination against pneumococcal and meningococcal organisms, use of hydroxyurea, chronic or exchange blood transfusion and continuous education and counseling [13,15,18]. This study was undertaken to evaluate the pattern and severity of acute painful crisis/VOC among children living with sickle cell anemia presenting to Lagos University Teaching Hospital Lagos with a view to understand the manifestations of SCA in this group of patients. It was hoped that the findings from this study will contribute to knowledge of the disease in children with sickle cell anemia in Nigeria.

Materials and Methods

This was a prospective cross-sectional study and involved patients who presented at the emergency room, or out-patient unit of the Pediatrics Department of the Lagos University Teaching Hospital, Idi-Araba Lagos with acute painful crisis over a nine month period (March 2015-November 2015). Vaso occlusive crisis is defined as occurrence of pain in the extremities, back, chests or any other parts of the body that led to hospital presentation and/or admission and could not be explained by any other disease entity except sickle cell disease [19]. The respondents were known SCA previously diagnosed in the hospital between ages 1-18 years, and regularly attend follow-up outpatient clinic of the paediatric sickle cell clinic. All SCA patients with confirmed chronic disease [CVA, CKD, chronic heart disease] and the newly diagnosed SCA children in acute painful crisis were excluded. The attending physicians interviewed the patients and questionnaires were completed. Ethical approval was obtained from Lagos University Teaching Hospital Health Research and Ethic Committee [LUTH HREC]. After an initial stabilization of the child, the details of the study were communicated to the parents/guardian and patients, consent and assent forms were completed by parents/guardians and patient aged 10-18years for subjects who satisfy the study criteria. A detailed clinical examination including the pain score was carried out in the Children Emergency room or on the wards and documented. The examination included anthropometric measures. The severity of pain in children aged 1-7years was assessed using the FLACC pain scale while Wong –Baker pain rating scale was used for children aged 8-18years. The FLACC pain scoring was done by 2 independent clinical investigators while the Wong –Baker was by the subject in pain. The children were managed as per the protocol of the institution [Children Emergency and the Haematology Units] in order to maintain the best clinical practice and to meet the patients` requirements.

Results

A total of one hundred and six children with VOC were enrolled in the study. The mean age of the study participants was 8±7 years and the age distribution showed that 36(34%) children each were in the 1-5years and 6-10years groups and 34(32%) were in the adolescent age group (11-18years). The male to female ratio was 1:1. Table 1 below shows the socio-demographic and anthropometric characteristics of the patients

Table 1: Demographic and anthropometric characteristics of study participants.

Characteristics of SCA Patients	N (%)
Age at enrollment: Median (IQR) years	8 (1-18)
1-5 years	36(34%)
6-10 years	36(34%)
11-18years	34(32%)
Gender	
Male	56(50%)
Female	56(50%)
BMI [Kg/m²]	
Low	95 (89.6%)
Normal	10(9.4%)
Overweight	1(1%)

Location of the vaso-occlusive crisis

Out of the 106 patients that presented in VOC, most of the respondents (51%) presented with pain in the extremities (upper and lower) while the rest presented with pain in the chest, back, abdomen and penile shaft. Only 2 patients presented with back pain, 21 patients presented with pain in the lower extremities only and 17 patients had pain in the upper extremities only. About one tenths of the respondents presented with pain in three or more regions of the body. The most frequent location of the pain was in the lower extremities and the least frequent site of pain was penile shaft (priapism). The location of pain is as shown in Table 2.

Table 2: Gross distribution of pain location.

Location of Pain	(%)
Extremities [Upper and Lower Limbs]	51.2
Abdomen	18.6
Chest	16.3
Back	13.4
Penile	0.6

Duration of pain

Fifty-nine (55.7%) children presented in the hospital within 2-4 days of the onset of VOC, while 4 (3.8%) of the patients presented after 7 days of onset of pain and only 14% presented within the first day of pain. Table 3 below shows the duration of pain prior to presentation in the hospital.

Table 3: Duration of Pain at Hospital Presentation.

Duration of Pain [in Days]	Frequency N (%)
≤1	14(13.2)
2-4	59(55.7%)
5-7	29(27.4%)
>7	4(3.8%)

Severity of pain

With respect to the severity of acute painful crisis, most of the

patients (63.2%) presented with moderate intensity of bone pain, while (32.1%) presented with severe pain intensity and only 4.7% presented in mild bone pain intensity. (Figure 1)

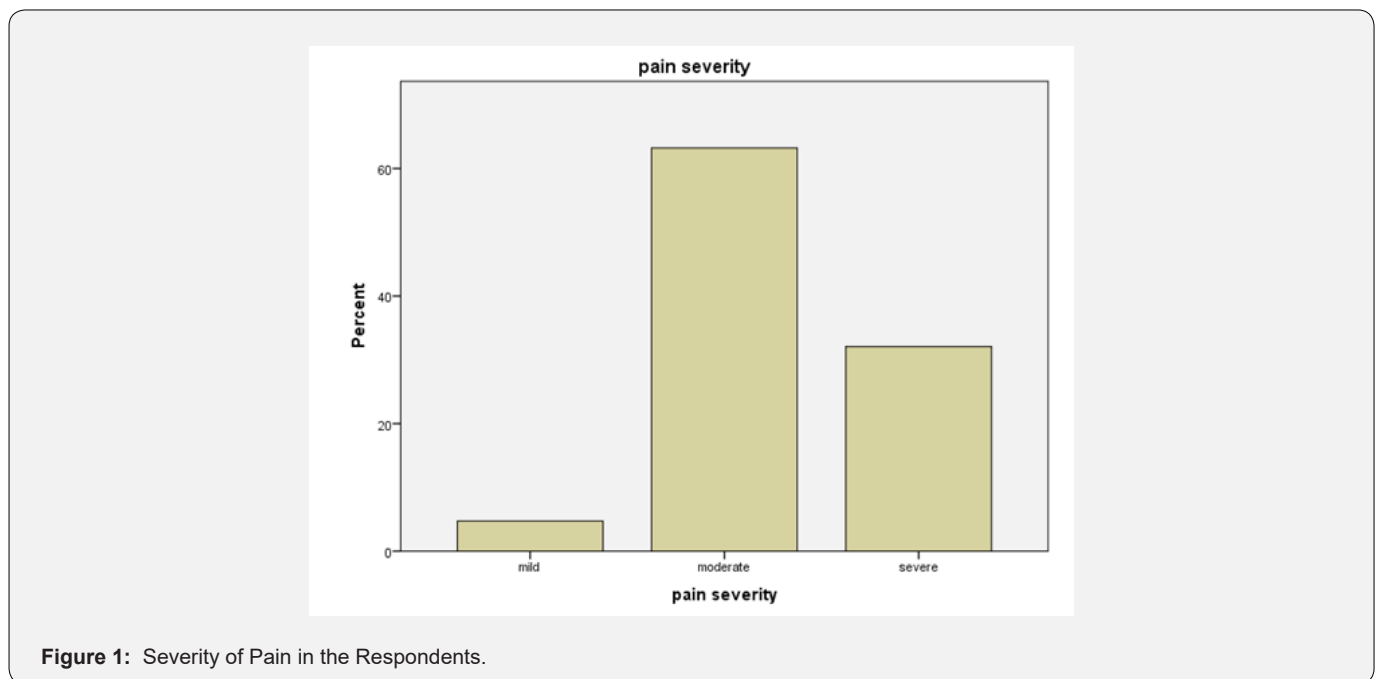


Figure 1: Severity of Pain in the Respondents.

Discussion

The index study is to evaluate the pattern and severity of acute painful crisis among children living with sickle cell anaemia using validated pain assessment tools; FLACC and Wong baker faces for children [1-7 years] and [8-18years] respectively. The anthropometric characteristics of the participants showed that most sickle cell anaemia patient are underweight, this could be as a result of inadequate growth hormone secretion, increased demand for nutrition and increased rate of metabolism, this finding is consistent with previous studies done by Odetunde IO & Andrea Zivot et al, [20,21] where most patients reviewed were found to be underweight. In this study, the commonest sites of pain were the extremities [upper and lower limbs] though pains in the lower limbs were commoner. This could be as a result of increased interruption of blood flow to the long bones because of

the sinusoidal circulation in these long bones. Other sites in order of frequency include abdomen, chest, back and penile shaft as previously documented [22]. These findings are consistent with previous studies done by Salman et al, [23] in Iraq, Jaiyesimi et al, [24] in Oman, and in another study with the paediatric age group in America and Europe [25] but contradicts the finding in some other previous studies that identified chest, abdomen and back as the commonest site of pain. The different findings in the publications explain the plausible variability in pain location in different individuals living with sickle cell anaemia. Also, most of the patients had pain in two or more sites, this is consistent with findings previously reported [26]. Most of the participants presented to the hospital within 2-4 days of onset of pain, this is because most of the patients and the parents/guardians take or give some form of analgesics prior to presentation in the hospital.

Moreover, vaso-occlusive crisis are initially mild at the onset and progressively increase in intensity over time. This finding is in concordance with previous works done by Jacob E et al. [27,28]. A significant proportion of the participants presented with moderate pain intensity. This is alluded to routine health education at sickle cell support groups on the use of analgesics for mild pain but when there is no improvement, they are to present in the hospital. However, some vaso-occlusive crises could be severe from onset and necessitate hospital presentation within the first day of pain. In this study, about one-third of the patients presented with severe pain. This could be responsible for presentations within the first 24 hours of the painful crisis. This finding is consistent with previous reports by Oshikoya et al. [29] in Nigeria, Abdo et al., in Jordan [30], Jacob & Neri et al. [27,31] in studies in the USA. This finding is in congruent with other reports evaluating the intensity of pain at presentation among children with sickle cell anaemia presenting in acute painful crisis despite the use of different pain assessment tools. [32-34]

Conclusion

In conclusion, the study further buttresses the findings that most SCA patients are underweight and the extremities are the commonest site of acute painful event. Most patients also present promptly to the hospital. The need for health education and prompt management of acute painful events in children with sickle cell anaemia will promote their total physical, psychosocial wellbeing of the children and their parent/caregivers.

Limitations

This study is characterized by some limitations; the proportion of the children who presented in acute painful crisis that required admission and the duration of the hospital stay with respect to resolution of pain or development of other clinical condition were not documented. This is a hospital-based study, a community study is needed to capture the patients not on routine follow up in our hospital.

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