Acute Painful Crisis In Sickle Cell Disease: Severity Assessment
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Citation

Abstract
The lack of an objective way of assessing the severity of sickle cell painful crises prompted this study. Adult sickle cell disease patients who presented to a tertiary hospital with acute painful episodes over a nine-month period were studied. The patients’ vital signs at presentation, the pain scale and the effect of pain on the patients’ daily activities were used in association with the doctor’s clinical assessment of severity. The effect of the pain on daily activities was closely associated with the doctors' and the patients' assessments (p=0.02). The systolic blood pressure and the respiratory rate were good indicators of the severity of pain. We suggest that the pain scale, the effect of pain on daily activities and the vital signs be used in conjunction with the doctor’s assessment in determining the severity of the painful crises.

INTRODUCTION
Pain is a universal phenomenon, which is unpleasant with wide variation in intensity, quality, duration and persistence (1). The acute painful crisis is a common manifestation of sickle cell disease, which accounts for over 60% of hospital admissions in any given year, yet its pathogenesis is poorly understood (2,3). The manifestations differ greatly between patients and in the same patient at different times. This has led to difficulties in standardizing management of these acute episodes. The lack of an objective way of assessing the severity of the painful crises has in turn made the choice of analgesics a difficult task. In this communication, we used both objective and subjective parameters to assess the severity of the painful episodes.

PATIENTS AND METHODS
The patients are those who presented to the day-care facility of a tertiary hospital with acute painful episodes over a nine-month period. This included adult patients who are over 15 years and comprised both HbSS and HbS+C patients. They were observed in the day-care unit for up to 8 hours following which they were discharged or admitted to the hospital if the pains were unabated. Patients who were admitted to the hospital through the accident & emergency unit of the hospital were also included in the study.

For each painful episode, the duration of the pain before presentation and the site(s) of the pain were recorded. The patient’s perceived cause of the pain was also recorded. The doctor assessed the severity of each episode as mild, moderate or severe. Similarly the patients assessed the pain as mild, moderate or severe. In addition the patients assessed the intensity of the pain on a scale of 1-10. A comparison of the assessments with the effect of the pain on daily activity was made. On presentation to the hospital the patient’s vital signs: pulse, respiration, blood pressure and temperature were recorded. The steady state haematocrit obtained by averaging at least five of the patient’s haematocrit when symptom-free was compared with the patient’s haematocrit at presentation. The duration of the patient’s stay in the hospital was noted.

The chi-square test of association was used to test the relationship between two nominal variables. Statistical significance level was taken as p<0.05.

RESULTS
Seventy-two patients were seen during the study period and consisted mainly of HbS (90%). There were 40 females (56%) and 32 males (44%), the male/female ratio of 1:1.3 is similar to a male/female ratio of 1:1.5 obtained from the general clinic population. Patients who presented within 24 hours of the onset of the pain accounted for 37.5% of the study population while 6.9% presented a week or more after the onset of the pain. The lower limb is the site most commonly affected by acute painful crisis (27.8%) multiple sites were involved in 45.8%.
Fifty-one percent of the patients classified their pain as moderately severe while 44.4% classified it as severe. The attending physician assessed 62.5% of the patients as having moderately severe pain, 20.8% as mild and 16.7% as severe pain. The attending physician assessed seventy percent of the patients who presented within 24 hours of the painful episode as moderately severe. Sixty-five percent of the patients were discharged home after observation in the day-care center (within 8 hours of presentation in the hospital), therefore not needing hospital admission.

The steady state haematocrit was not significantly different from the haematocrit at presentation \((p=0.62)\) but the steady state haematocrit showed better association with the patient's assessment of the pain \((p=0.02)\) than the haematocrit at presentation \((p=0.7)\). There is close association between doctors assessment and the patients use of the pain scale to asses the pain \((p=0.009)\). Temperature was closely associated with the patients' use of the pain scale \((p=0.02)\). The effect of the pain on daily activities was closely associated with both the doctors' assessment \((p=0.02)\) and the patients' assessment of the pain \((p=0.02)\). The assessment of the pain by the doctor show close association with systolic blood pressure \((p=0.01)\) and the respiratory rate \((p=0.01)\).

**DISCUSSION**

This study has shown that sickle cell disease patients in this environment do not present early to the clinic when in painful crises, less than forty per cent of the patients presented within 24 hours of the onset of the pain, in comparison to Jamaican patients in whom more than half of the patients present within 24 hours \((2\%)\). Similarly only 35% of our patients and 39% of Jamaican patients required admission for painful crises, this contrast sharply with 92% of sickle cell disease patients in Britain who needed hospital admission for painful crises \((2\%)\). Reasons for this may include the colder climate that may result in a more severe and frequent crises. Easy assess to health care will also encourage more patients to present early when in pain.

Epidemiological and prevalence studies reveal that women have greater and more pain than men \((2\%)\), with girls reporting more pain than boys after surgery \((2\%)\). Women have also been found to score the intensity of their pain significantly higher than men \((2\%)\). On the contrary, gender does not appear to influence the painful crises in sickle cell disease patients. This study shows that the sex ratio is not significantly different from that of the general patient population and that the intensity of pain reported by the patients is unrelated to gender. This finding is not peculiar to this study population because a similar finding was noted in Jamaican patients \((2\%)\). Trunk involvement was found to increase with age \((2\%)\) presumably because haemopoiesis occurs more in the flat bone with advancement in age. This study showed that about half of the patients complained of multiple site involvement, with the long bones being more affected than the flat bones.

Ability to objectively assess the painful episodes in these patients was difficult to achieve. This is because, the assessment of the pain by the patients differ from the doctors' assessment of the pain. The use of a pain scale and the effect of the pain on the patient's daily activity were better tools than classifying the pain as mild, moderate or severe by either the patient or the doctor. The effect of the pain on the patient's daily activity appears to be a better tool since there is close association between it not only with the doctors assessment of the pain but also with the patients assessment of the pain. Fever is accepted as a common feature of uncomplicated painful crises and often resolves without the use of antibiotics \((0\%)\). Malaria is endemic in this environment and so its effect is a confounding factor that needs to be considered.

The systolic blood pressure and respiratory rates which are physiological measures were found to be more closely associated with the doctors' assessment of the painful crises than the patients' assessment. The use of a pain scale and the effect of the pain on daily activities to assess the pain take the mind of the patient temporarily off the pain and so are better tools in assessing the pain than classifying the intensity of the pain as mild, moderate or severe by the patient. We would therefore suggest that to assess the severity of the patients' pain, both subjective and objective criteria be employed. The subjective criteria would include; the use of a pain scale, the effect of the pain on daily activities and the doctors assessment while the objective criteria would include the systolic blood pressure and the respiratory rate.

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