

Management of acute painful crises in sickle cell disease

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Summary Pain is a common mode of manifestation of sickle cell disease (SCD) but there is limited information on pain management in this disorder. This study examines the use of opioids and non-opioid analgesia in the management of painful crisis in adult SCD patients; the routine use of antimalarials and antibiotics as adjunct therapy was also examined. A total of 87% of the patients had had a form of analgesics before presentation, 20% of which had parenteral analgesia. Ten per cent had not used any form of medication while another 10% used non-steroidal anti-inflammatory drugs. When asked, 59% of the patients desired oral non-opioid analgesics while 31% were not concerned about the type of analgesic given. Only 8% requested opioids. Hospital admission was not necessary in 65% of the patients; they were observed in the day-care unit and allowed home within 24 h. Sixty per cent did not have a test for malaria; 66% of those who had the test performed were negative, 35% of those whose thick film for malaria was negative had antimalarials prescribed. Only five patients (7%) were febrile at presentation. Thirty-four per cent had antibiotics prescribed, a third of these parenterally. Thirty-nine per cent had no fever but received antibiotics.

Keywords Painful crisis, opioids, analgesia, sickle cell disease

Introduction

Pain is a common mode of presentation in patients with sickle cell disease (SCD) but there is considerable variability in the way SCD pain is managed. The standard treatment protocol for painful episodes has remained bed-rest, hydration and analgesia. There are a variety of analgesics to choose from, both opioids and non-opioids, which could either be given orally or parenterally depending on the severity of the pain. One of the factors contributing to poor pain management is conflicting perceptions between patients, their families and health-care professionals about pain that is reported and analgesia that is required (Stinson & Naser, 2003). Pain management guidelines have recently been published in an effort to overcome barriers in the assessment and management of pain related to SCD (Okpala & Tawil, 2002; Rees *et al.*, 2003), the effect of which is yet to be evaluated. In this study, we have looked at the use of drugs in the management of SCD patients thus highlighting the problems encountered in such a setting.

Patients and methods

The patients are those who presented to the day-care facility of a tertiary hospital with acute painful episodes over a 9-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 a maximum of 8 h following which they were either discharged home or admitted to the hospital if the pains were unabated. Patients who were admitted to the hospital through the 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 duration of hospital stay also noted. Medications used before presentation to the hospital and the choice of analgesic for the pain was inquired from the patient. The doctor assessed the severity of each episode as mild, moderate or severe. Analgesic was prescribed based on the doctor's assessment. Antibiotics and/or antimalarials were prescribed as deemed fit by the doctor. Patients whose pain was adjudged to require bed rest and/or fluid therapy were offered these while on observation in the day care while patients with mild pain not requiring parenteral medication were allowed home after having been attended by the doctor. Those who required parenteral medication subse-

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quently had oral medication with additional parenteral medication for breakthrough pains. Information was obtained by the use of a semi-structured questionnaire and from the doctors' documentation in the case-files.

Results

There were 40 females and 32 males. Sixty-two per cent of the patients were in a tertiary institution or had completed a tertiary education while only 1% had at least a primary education.

A total of 87% of the patients had had a form of analgesics before presentation, 20% of which had parenteral analgesia. Ten per cent had not used any form of medication while another 10% used a non-steroidal anti-inflammatory drug (NSAID). When asked, 59% of the patients desired that oral non-opioid analgesics be prescribed while 31% were not bothered about what analgesic was given. Only 8% requested opioids.

A total of 60% did not have a test for malaria (thick blood film) carried out; 66% of those who had the test performed were negative. Forty-eight per cent of the study population and 35% of those in whom thick film for malaria was negative had antimalarials prescribed. Only five patients (7%) were febrile at presentation (of the five, three did not have a test for malaria, while one had a positive result and the other a negative result). Thirty-four per cent had antibiotics prescribed, a third of these parenterally. Thirty-nine per cent had no fever but received antibiotics.

A total of 65% of the patients did not require hospital admission but were observed in the day-care unit and allowed home within 24 h. Only 17% required hospital admission for more than a week.

Discussion

In non-endemic areas, the diagnosis of clinical malaria may be made based on fever and a positive blood film. However, in areas of high endemicity, asymptomatic parasitaemia is very common (Schellenberg *et al.*, 1994). It is therefore necessary to correlate symptoms of malaria with parasitaemia to avoid overdiagnosis. It appeared that neither parasitaemia nor symptoms of malaria were considered before antimalarials were given to the patients as only 7% of the patients were febrile at presentation and 60% did not have blood smears for malaria testing yet 48% had antimalarials prescribed. Despite the fact that infections are an accepted cause of painful episodes in SCD, it is necessary to confirm malaria as the precipitating factor with a microscopic examination of blood smears

which is the 'gold standard' for malaria diagnosis (Fernando, Karunaweera & Fernando, 2004; Soto *et al.*, 2004) and fortunately still affordable in a developing economy. It also appeared that not much reliance is placed on the result of the blood smear as 35% of those with a negative smear still had antimalarials. The use of antibiotics also followed the same trend because 34% of the patients also had antibiotics despite the fact that only 7% were febrile and a third had it parenterally. It is therefore necessary to confirm infection as the precipitating cause of the vaso-occlusive crisis before commencing treatment.

One of the factors contributing to poor pain management is conflicting perceptions between patients, their families and healthcare professionals about pain that is reported and analgesia that is required (Stinson & Naser, 2003) and this stems from concerns about dependence. Although there had been observations of abuse of narcotics among a few of our patients, this report does not show much abuse of narcotics but rather an attempt to anticipate pain and avoid hospital admission (87% had received some form of medication before presenting in the hospital). This underscores the observation of Elander *et al.* (2003) that an SCD patients' pain coping may be perceived as analgesic dependence (Merle, Thieffin & Czernichow, 2004).

Gastrointestinal lesions are accepted complications of the use of NSAID (Maende *et al.*, 1998; Richy *et al.*, 2004). Endoscopic and gastric acid studies have also shown that mucosal lesions are commoner in SCD patients than in sex- and age-matched controls (Lee *et al.*, 1989; Maende *et al.*, 1998). The use of NSAID either by the patients before presenting to the physician or by the physician should be especially discouraged because sickle cell pain is a recurrent pain which would almost always require the use of medication before abating.

Pain control with the use of intramuscular analgesia is believed to be unsatisfactory in sickle cell patients, which is the reason for the use of patient-controlled analgesia (PCA) in vaso-occlusive crisis in some centres (Holbrook, 1990; McPherson *et al.*, 1990). The use of PCA is uncommon in our setting yet adequate analgesia is achieved in the majority of our patients with 65% of patients not requiring hospitalization while in one study patients' pain scores decreased by only about 30% after 3 days of therapy with PCA (McPherson *et al.*, 1990). PCA may not be without adverse effects or major problems as previously thought (McPherson *et al.*, 1990), as some patients have identified some of the problems of PCA as ineffective analgesic regimens, analgesic side effects and technical shortcomings as well as reduced direct nurse contact (Johnson, 2003). The total dose of intravenous

narcotic therapy was found to be greater in patients who had PCA than in patients who had nurse-administered analgesia (Holbrook, 1990) despite the fact that time in the hospital, time until pain relief and duration of parenteral narcotic therapy were similar in both groups (Holbrook, 1990; Gonzalez *et al.*, 1991). The choice of opioids analgesic, its route of administration, dose and frequency of administration should therefore be individualized.

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