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CLINICAL PRACTICE

Pain Management in Adult Acute Sickle Cell Pain Crisis: A Viewpoint

Chagriner la direction dans la crise de douleur de cellule de faucille adulte: un point de Vue .

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ABSTRACT

BACKGROUND: The acute pain crisis of sickle cell disease is inadequately treated in many countries.

OBJECTIVE: To present a simple protocol that controls acute pain in most adult patients within 72 hours, based on our experience in an area where sickle cell disease is highly prevalent.

METHODS: Patients aged 14 years and above with sickle cell disease presenting with pain crisis are treated initially in the Emergency Room. Those responding adequately are discharged home on oral analgesics while those with persisting pain after 6 hours are treated further, for up to 72 hours, in an Observation Unit attached to the Emergency Room. Narcotic analgesics are administered regularly for the first 24 hours.

RESULTS:Using this protocol, acute pain crisis could be terminated or controlled in over $80\,\%$ of patients within 72 hours.

CONCLUSION: Regular intravenous narcotic analgesia for the initial 24 hours, supplemented by oral analgesia, is useful for adult patients with severe acute sickle cell pain crisis when the pain is inadequately controlled by initial Emergency Room treatment. There may be gender differences in pain perception and response in acute sickle cell pain crisis. WAJM 2007; 26(3): 179 – 182.

Keywords: Sickle cell disease, acute pain crisis, pain control and response.

RESUMÉ

Contexte: La crise de douleur aiguë de maladie de cellule de faucille est inadéquatement traitée dans beaucoup de pays.

Objectif: Pour présenter un protocole simple qui contrôle la douleur aiguë dans la plupart des malades adultes dans 72 heures, basé notre expérience dans un secteur où une maladie de cellule de faucille est extrêmement courante.

Methodes: ont vieilli 14 années et au-dessus avec présenter de maladie de cellule de faucille avec la crise de douleur sont traités au début dans la Pièce d'Urgence. Ces répondre est suffisamment déchargé à la maison sur les analgésiques orales pendant que ceux-là avec persister de douleur après 6 heures est plus traitée, pour augmenter à 72 heures, dans une Unité d'Observation a attaché à la Pièce d'Urgence. Les analgésiques de narcotique sont régulièrement administrées pour le premier 24 heures.

Resultats: Utilisant ce protocole, cette crise de douleur aiguë pourrait être terminée ou pourrait être contrôlée dans pardessus 80% de malades dans 72 heures.

La Conclusion: analgésie de narcotique intraveineuse Régulière pour l'initiale 24 heures, complétées parl'analgésie orale, sont utiles pour les malades adultes avec la crise de douleur de cellule de faucille aiguë sévère quand la douleur est inadéquatement contrôlée par le traitement de Pièce d'Urgence initial. Il peut y avoir les différences de sexe dans la perception de douleur et la réponse dans la crise de douleur de cellule de faucille aiguë. WAJM 2007; 26(3): 179 – 182.

Mots: la crise de douleur aiguë, la douleur contrôle et la réponse.

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Abbreviations: ER, Emergency Room; HbSS, Sickle Cell State; iv, intravenous; NSAID, nonsteroidal antiflammatory drugs; SCD, Sickle Cell Disease; SU, Stabilization Unit

INTRODUCTION

Sickle cell disease (SCD) which includes the homozygous sickle cell state (HbSS) and the double heterozygous condition with haemoglobin C disease, (HbSC), is prevalent in many tropical countries, not only in Africa, but also in India, the Middle East, USA, and South America. It has several forms and haplotypes with variability in clinical presentation and severity¹. However, its clinical hallmark, the acute pain crisis, caused by vaso-occlusion of small blood vessels by haemoglobin S-rich red cells which sickle as the haemoglobin polymerises on deoxygenation, is present in all forms. It can affect any or multiple parts of the body but especially the extremities, back, chest, and abdomen. Pain control improves quality of life, and life expectancy; more symptomatic SCD is associated with increased mortality 2.

SCD is prevalent in Eastern Saudi Arabia where the responsible gene occurs in 25-30% of the population ⁴. Consequently clinical SCD is very common and we treat over 1000 cases annually in our Emergency Room (ER) and attached Stabilization Unit (SU), serving an 85,000 health care population of Saudi Arabian Oil Company (Saudi ARAMCO) ⁵. We have tried various modes of treatment for this condition over the years, and what follows is drawn from this experience.

A generalist approach is taken; hence advances and treatment modalities available only to specialist centres including hydroxyurea therapy, blood and exchange blood transfusion ^{6,7} are not discussed. Since SCD is clinically relatively 'mild' in Eastern Saudi Arabia compared with the western part of the country or West Africa ⁸⁻¹⁰ we modified our treatment according to the local pattern of the disease.

PATIENTS AND METHODS

Adult SCD patients who present to the ER, in painful crisis are assessed for pain and treated initially by ER Physicians. Pain is scored on a scale of 0-5, using an accepted scale such as the pain faces of Wong-Baker (Fig 1)¹¹, or our own custom designed colour code (Fig 1). Treatment consists of intravenous re-

Table 1: The Protocol for Treatment of SCD Pain Crisis in the ER Observation Unit.

- Oxygen: when necessary by nasal cannula to keep oxygen saturation by pulse oximeter (SPO2) = 95%
- Adequate hydration: (avoid parenteral over-hydration which may precipitate Acute Chest Syndrome, ACS)

Oral fluids: 2-3 litres of water or fruit juice daily, whenever possible or iv 0.45% saline in 5% dextrose at 1-1.5ml/kg body weight /hr ie 75-125ml/hr for first 24 hrs, then review fluid intake. (Avoid 'normal' or 0.9% saline which is actually hypertonic as it contains 154mmol Na+/L; it may contribute to ACS).

Initial Pain control with i/v morphine 5
 7.5mg Q4 hrs REGULARLY (Not when necessary) for first 24 hours, then review and change to 'on demand' basis (eg 5mg prn Q 6hrs) when pain is controlled; REASSESS PATIENT FREQUENTLY UNTIL PAIN CONTROL IS ACHIEVED (Use both patient's report and nurses' Pain score sheet to assess pain control).

Add i/v promethazine 12.5-25mg Q8hrs to decrease morphine induced nausea and vomiting, and also potentiate its effects.

Also start concomitant ORAL ANALGESIC eg Paracetamol Q4hrs or one of the following: Ibuprofen 400-600mg Q 8 hrs or Diclofenac 50mg Q 8hrs or Naproxen 250-500mg Q 8hrs or Any other available analgesic or non steroid anti-inflammatory drug, (NSAID)

4. Re-evaluate patients 6-hourly or more often if indicated, and adjust analgesia until pain is controlled. Assess pain relief against patient's report using pain chart untilain is controlled. Assess pain relief against patients' reports, using pain charts.

Intravenous (IV) Ketorolac, 30-60mg, repeated if necessary, 6hourly; iv Morphine 2-5mg, half-hourly until pain is controlled or iv Pethidine (Demerol), 50-100mg, 4hourly when necessary. Repeated small doses of narcotics minimize the risk of respiratory depression with over dosage.

Patients who respond adequately are discharged home on oral analysics of their choice within those available in our formulary, while those with significant persisting pain, are admitted to the SU. However, our ultimate indicator

of adequate pain relief was the proportion of patients discharged home or transferred to in-patient care.

RESULTS

Using this protocol, acute pain crisis could be terminated or controlled enough for patients to go home in over 80% of patients within 72 hours. About half of patients usually went home after initial ER treatment while the other half required further treatment and observation. Regular intravenous narcotic analgesia was more effective than intermittent or 'on demand' dosing with a higher discharge rate (83% vs 71%, P<0.05)¹³. A minority of patients, about 15-20%, had more prolonged pain requiring in-patient treatment.

Male patients seemed to fare worse than females, with more recurrent, persistent and prolonged pain crises, leading to more hospitalizations. More males than females required further inpatient treatment (Table 2). They also had more frequent crises¹⁴.

Morphine was more effective than pethidine whose 2-3 hours duration of action was found to be too brief in our earlier series (1995-97) to be practically useful. Intramuscular administration of both of narcotic and non-narcotic analgesics gave inferior pain control and was not liked by patients. In 100 patients, using oral NSAIDs, Ibuprofen (31%), diclofenac and mefenamic acid 17% each, and naproxen (25%), were the most popular . Only 1% preferred paraccetamol. A few patients requested diclofenac specifically because of previous experience of its efficacy.

Table 2: Outcome of SCD Pain Crisis
Treatment.

Treatment.		
Period*	1995 - 97	2000 - 02
Discharged hon	ne (%)	
	71	83
No of Patients	305	849
Males	151	422
Females	154	427
Males hospitali	zed <i>N</i> (%)	
	53(35.1)	98(23.2)
Females hospita	alized $N(\%)$	
_	36 (23.4)	47 (11)

^{*1995-97} On demand Pain Treatment; 2000 - 02 Regular Pain Treatment.

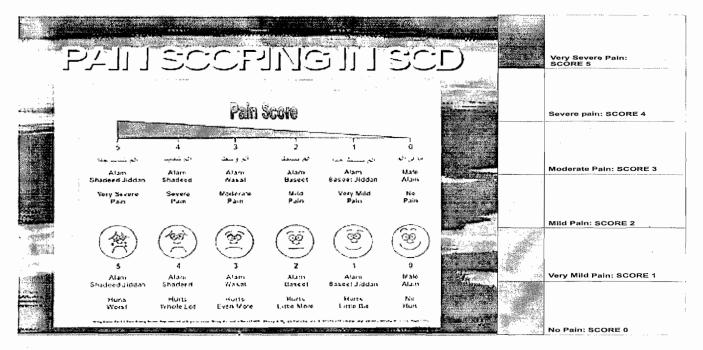


Figure 1: Ahhc Internal Medicine Unit: Pain Severity in Sickle Cell Disease. Pain is scored on a scale of 0-5, using an accepted

DISCUSSION

Sickle cell disease symptoms including pain are indicators of the tissue injury caused by the vaso-occlusion of sickling, hence their association with increased mortality ². Pain control is therefore important. On demand analgesia systems work, but several factors often interfere with the schedules for this mode of drug administration³. Patient controlled analgesia systems are probably the best way to administer analgesia in this condition, but they are expensive in terms of the required equipment, staff training and dedication.

Morphine is more effective, cheaper and longer lasting in effect than pethidine which is associated with more addiction and other adverse effects¹⁵⁻¹⁷. Intramuscular Diclofenac is very painful, and frequent intramuscular injections of painful drugs are not recommended for long-lasting pain like that of SCD¹⁸. Ketorolac is very expensive: a standard 30mg dose is less effective and costs 10 times as much as 5mg morphine from a multi dose vial; it has also not lived up to its narcotic-sparing promise or expectations^{19, 20}. Several different oral analgesics should be available in treating centres because of variable patient response to them.

Males may require up to 40% more narcotic analgesic dosage than females of similar body weight, to get the same relief ^{21,22}. Gender differences in pain response require further investigation. ¹⁴ Treatment should therefore be individualised for SCD patients in crisis because of their variable clinical presentation, course and response to medications. Their need for frequent pain medication should be recognized by all concerned, so that appropriate treatment can be given, and their live burdens eased.

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