

North West London Haemoglobinopathy  
Managed Clinical Network

**Guidelines for the Management of Acute Pain in adult  
patients with sickle cell disease**

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### **1. Background**

Sickle cell disease (SCD) is a hereditary disease affecting mostly people of Afro-Caribbean/African, Mediterranean, Middle Eastern and Asian descent. Patients with SCD experience painful episodes of vaso-occlusive crisis caused by deformed sickle shaped red blood cells which can lead to tissue infarction. This leads to various levels of pain with 90% of admissions occurring with severe pain due to a vaso-occlusive crisis. If the pain is left untreated for a prolonged period it can lead to a multitude of complications.

The recommended treatment for severe pain during these episodes are strong opioids given either by parenteral route (by injection or infusion) or orally; This is achieved by administering regular opioids with rescue doses for breakthrough pain or using a patient controlled analgesia (PCA) method. Subcutaneous (s/c) route for administering parenteral opioids is preferred rather than the intravenous route due to poor venous access in this patient population and to enable prompt initiation of treatment.

This guideline will guide the clinical teams within the local North West London (NWL) Managed Clinical Network (MCN) for Haemoglobinopathies to manage painful crises effectively.

Please use these guidelines in conjunction with the latest NICE guidance (June 2012, reviewed, April 2014) on acute pain management for patients suffering an acute painful episode.

### **2. Aim**

The aim of this guideline is to enable appropriate, prompt, adequate and safe analgesia to be administered to SCD patients during a vaso-occlusive crisis and pain relief within two hours of presentation. Vaso-occlusive crisis should be treated as an emergency situation and preferably managed by a specialist team with expertise in this condition.

### **3. Scope of practice**

This guideline is solely for the management of acute painful crisis in an adult SCD patient. For good pain management it is preferable that the haematology team caring for the patient manage the individual's acute pain rather than the Trust's acute pain team to provide prompt management and continuity of care. For chronic pain management it is advisable to contact the Trust's pain team for advice.

All patients under the care of the North West London Network should have an individual analgesia protocol for pain management which should be reviewed at least annually. Individual analgesia protocols potentially communicate information about management in a timely manner aiming for the first dose of analgesia to be administered within 30 minutes of presentation and to receive significant reduction in pain score within 2 hours of presentation.

Options for opioid administration:

- 1) subcutaneous (S/C) route as required (PRN) 2 hourly **or**
- 2) patient controlled analgesia (PCA).

**Neither option's should be used in combination on the drug chart due to increased risk of side-effects.**

- 3) Some Trusts may use intravenous opioids according to local policy as bolus doses or PCA.

### 3.1 Pain assessment

a) To assess the patient's pain, at first presentation, the patient must be asked the following questions by both medical and nursing teams:

- History; cause, previous treatment and drug history
- Pain score according to local pain assessment tool
- Site of pain and type: acute or chronic
- Other symptoms and physical examination; general, vital signs

b) Document all findings in medical notes.

c) A detailed pain assessment should be recorded before and after administration of pain relief.

d) Assessment of pain is a vital element for diagnosis and for the evaluation of pain management.

e) Only the person experiencing the pain knows its nature, intensity and location. Observe for verbal and non-verbal signs of pain during assessment. Regard the patient as an expert in their condition by listening to them and discuss treatment plans with them at all times.

### Guidelines for pain assessment

- Pain assessment should be completed every 15-30 minutes for the first two hours of presentation and 4 hourly thereafter if stable. Further assessment should be made as indicated by the patient's condition and use a locally approved pain score as recommended by the Trust pain team.
- If parenteral opiate dose is increased the pain assessment regime should be commenced again as appropriate.
- If prescribed parenteral opioids are not controlling the patient's pain within the emergency acute setting inform the haematology team as soon as possible. Uncontrolled pain can be indicated by the frequency of opiate PRN injections or number of PCA boluses (with good presses) versus demanded doses on the PCA. Educate the patient to be able to communicate accurately their level of pain and requests for pain relief before increasing parenteral opiates.
- After reassessment, within the first 30 minutes of presenting, if the patient remains in moderate or severe pain (i.e. pain score  $\geq 3$  of NEWS score 1-4) give a second dose reduced by 50% of the patient's initial opiate dosage. NICE uses a score  $>7$  for severe pain and 4-7 for moderate pain (0-10 scale).
- Consider other additional methods of pain relief such as massage, positioning, psychological interventions (distraction and cognitive behavioural therapy), topical analgesia or apply warm heat to affected area. **Do not use either corticosteroids or transcutaneous electrical nerve stimulation (TEN's) for an acute crisis as there is limited evidence proving their benefits.** TEN's is only recommended for chronic localised pain.
- If pain is reported by the patient as atypical explore an alternative diagnosis such as abdominal pain and jaundice, priapism or infection. (See local clinical guidelines for further advice).

- Nitrous oxide/oxygen 50/50 (Entonox) mix for pain control should be limited to no more than 30-60 minutes within the ambulance and emergency setting. Use with caution in patients with hypoxia; respiratory rate >24 or <12 per minute/ oxygen (O<sub>2</sub>) saturations <92% or acute chest syndrome. Not to be continued long-term because of the risk of megaloblastic anaemia and neuropathy.

#### **4. Eligibility and special precautions for opioid administration:**

##### **Eligibility**

The patient must be:

- Conscious, appropriately responsive and able to obey commands
- Respiratory rate >10/min and O<sub>2</sub> saturations on room air >92%
- Or O<sub>2</sub> saturations >95% on oxygen therapy.

For PCA administration:

- Verbal consent and able to operate the PCA handset effectively.
- Patients agrees verbally not to leave the ward whilst on PCA

If a female SCD patient presents in pregnancy it is safe to use parenteral opioids during the painful crisis. Avoid NSAIDs, tramadol, pregablin and gabapetin due to possible risks of foetal abnormalities.

##### **Special Precautions:**

- Patient is confused, unconscious, and unable to obey commands or has a high level of anxiety.
- Opioids for parenteral use that may be contra-indicated/caution in any specific patients e.g. allergy or respiratory disease; sleep apnoea, acute asthma attack or severe COPD. Seek respiratory medical advice if opioids are required.
- Patient refuses or unable to use PCA or parenteral/ s/c injections

- Use parenteral opioids with caution in patients with moderate to severe renal or liver impairment; discuss with the ward pharmacy and the pain team for further advice.

### 5. Use of Parenteral Opioids

Pain severity should be managed according to the World Health Organisation (WHO) analgesia ladder.

If the patient presents with pain when self-administered oral pain relief has not been effective use a multimodal approach of oral medications with NSAIDs (unless contraindicated) with suitable parenteral opioids. Follow the patient's own individual analgesia protocol for their pain management, rather than what the patient states. If the patient is requiring opiates for more than five to seven days then consider referral to the pain team and assess for other possible underlying complications.

For patients in adolescent transition continue to follow the local children's guidelines. Note at specialist centres oral oramorph or severdol is 1<sup>st</sup> line and 2<sup>nd</sup> line respectively via an IV PCA (if in paediatrics). Sub/cut (Adults).

Note that the use of strong opioids for persistent pain over seven days maybe associated with additional side effects such as hyperalgesia.

#### 5.1 Choice of parenteral opioids (bolus doses):

Diamorphine Dose Range 2.5-20mg s/c 2 hourly PRN

Morphine Sulphate Dose Range 2.5-30mg s/c 2 hourly PRN

Oxynorm Dose Range 2.5-20mg s/c 2 hourly PRN

Pethidine 50-100mg IM 2-4 hourly PRN is only to be administered if the patient has proven severe allergy/side-effects to all other forms of opioid analgesia. There is proven evidence that there is an increased risk of fits and dependence.

Tramadol or codeine phosphate are not recommended in acute painful crisis in combination with parenteral opiates.

All parenteral (s/c/parenteral) opioids should be prescribed as PRN two hourly with a dosage range according to their individual protocol. Discuss with the patient's ward consultant/SPR/CNS before prescribing increased dosages.

### 5.2 Adjuvant therapy

Consider prescribing the following drugs:

- Paracetamol 1 gram PO QDS or IV TDS. Maximum 4 grams in 24 hours (if below 50kg use 750mg)
- Ibuprofen 200-400mg QDS or Naproxen 500mg TDS
- Naloxone 200-400mcg s/c or IV PRN or mixed with opioid infusion (See respiratory function 7.1)
- Cyclizine 50mg PO, IM or IV TDS PRN (See nausea and vomiting 7.3)
- Senna 15mg PO Nocte regular (See constipation 7.5)
- Lactulose 10mls PO BD regular (See constipation 7.5)
- Hydroxyzine 25-50mg orally PRN TDS or other locally used anti-histamines (See Pruritus 7.4)
- Patients having repeated prolonged crises may benefit from the early addition of Gabapetin/ Pregabalin.

### 6 Patient controlled analgesia (PCA)

A PCA is a mechanical device which allows the patient to regulate the amount of analgesia received from their assessment of the magnitude of their pain. PCA aims to provide a safe and effective analgesic regime that is applicable to the individual and allows them to play an active role in the management of their pain. A PCA pump may provide a constant continuous (background) infusion rate plus patient controlled boluses. PCA avoids the peaks and troughs in blood plasma levels that are associated with injections if the continuous infusion rate is available. It is recommended avoiding

repeated painful injections as PRN due to the risk of local infection, tissue damage and the unpredictable rate of absorption with injections.

### 6.1 Starting the PCA

The PCA must be prescribed on the drug chart **and not prescribed with other opioids**, either strong or weak.

Patients must be instructed how to use the PCA to manage their pain by nursing staff trained in the use of PCA.

### 6.2 Subcutaneous PCA regimes

Setting up values (use local guidelines for dosing and preparation)

(Contact Clinical Nurse Specialists at specialist centres for regimes)

Regime	Diamorphine	Morphine	Oxycodone	Fentanyl
Amount				
Concentration				
Background Continuous Infusion rate				
Lockout time				
PCA bolus dose range				

Prescribe and start S/C injections initially until the PCA bag is ready to start. Once the PCA has started then stop sub/cut injections as they cannot be used in combination.

A background infusion may be suitable in patients who are long-term users who show high opioid requirements when on S/C opioids and complain of waking in the night with severe pain. Use background rate with caution in opiate naïve patients due to risk of respiratory depression and drowsiness. The background/ continuous infusion rate should start at a low dose and alter the bolus doses as appropriate.

### 6.3 Procedure (guided by local policy)

- All patients starting on PCA/parenteral opioids should have vital signs documented on observation chart, pain score obtained, nausea score, sedation score and respiratory rate prior to commencing PCA.

- Explain the procedure to the patient
- Check the drug prescription chart and prepare the solution by two registered nurses who are intravenous medication trained.
- Programme the pump in accordance with the prescription
- The PCA infusion, the prescription and the PCA programme must be checked by two registered nurses (Intravenous medication trained) one of whom has completed the PCA in-house training.
- Identify the patient, checking the patient's hospital number on the ID bracelet and the drug chart match.
- Use aseptic non-touch technique (ANTT) guidelines (follow infection control guidelines) to insert /set up the medication and insert the butterfly needle into the subcutaneous tissue and attach the PCA to the patient. Once needle is inserted a bio-occlusive dressing should be applied and dated. Clearly label the PCA giving set with date, time and drug.
- Press the start button, sign the drug chart and re-check that the patient understands how to use the PCA.
- Commence care and observation monitoring as appropriate.

**If using intravenous PCA follow local policy/ guidelines.**

### **6.4 Subcutaneous (S/C) needle insertion, use and care for PCA and injections**

Patients should have flow-safe winged infusion set sub/cut butterfly needle (e.g. Smith's medical) inserted as below. The date of the insertion should be recorded within nursing documentation and needle should be removed at 72 hours. When the insertion site becomes painful, red or after 3rd bag of PCA infusion set is change every 24 hours.

Placement and insertion of the needle should be in the following areas only; can also be used for frequent s/c injections. In order of preference:

- Anterior abdominal wall
- Upper thigh

- Deltoid area (upper aspect of the non-dominant arm if possible)

The sub/cut insertion site must be observed regularly (at least every shift) for signs of infection, abscess formation or leakage.

Only the opioid is to be delivered through the butterfly needle.

A separate cannula should be used if any other drugs are prescribed via sub/cut or Intravenous route administration.

### 7. Care and monitoring of the patient

Patients must be advised not to leave the ward while on the PCA or parenteral opioids. If the patient repeatedly leaves the ward despite warnings the continuation of this treatment should be discussed with the haematology consultant/team.

#### 7.1 Respiratory function

Assessment of the patient's respiratory function should begin with an overview of the patient's general appearance and by measuring their respiratory rate and oxygen saturations on room air. Normal involuntary respirations are regular, effortless and quiet. Irregularities in breathing may relate to rate, rhythm or volume. **If the patient's oxygen saturations and respiratory rate are stable there is little evidence available that oxygen therapy for a painful crisis is of any benefit.** Therefore only administer for a patient with acute chest syndrome, unknown hypoxia or chronic lung changes on an individual basis (normal respiratory rate range 12-20 per minute and oxygen saturations, normal range over 94% on air).

#### Respiratory complications

If the respiratory rate falls to below 10/minute or oxygen saturations on room air below 92% the PCA should be stopped. Treat patient with a potential risk of respiratory depression and inform doctor immediately.

An increase in respiratory rate of >24/min may signify acute chest syndrome and also warrants urgent medical assessment.

If respiratory depression is triggered the following should be done:

- Stop the PCA/inject able parenteral opioids

- Stimulate the patient
- Administer oxygen therapy via a re-breath mask for up to 15/litre until O2 sats are greater than 96%
- Summon emergency assistance
- Commence pulse oximetry, blood pressure, pulse and respiratory rate monitoring every 15 minutes.
- Naloxone must be given with great caution to patients who have received longer-term opioid/opiate treatment for pain control or who are physically dependent on opioids/opiates. Naloxone 100-200 micrograms must be available and prescribed. Naloxone can be given intravenously or sub/cut every 2-3 minutes until the patient is breathing normally, if response is inadequate give a further 100mcg. Maximum total dose is 1.5-3mcg/kg; however further doses may be given if respiratory function deteriorates on the advice of the medical team or emergency team. If Naloxone is given the medical team must be informed and observations made every 15 minutes in the first instance for one hour. Be aware naloxone will reverse the analgesic affect quickly due to a short half-life and patient will be in pain again quickly. Therefore use a reduced sub/cut parenteral opiate dose by 50% and monitor closely.
- A bag and mask must be available to ventilate the patient if necessary.

### 7.2 Sedation

The cause of an abnormal decrease of consciousness may be related to unintentional overdose of opioids. However in SCD there may be other associated causes e.g. stroke or meningitis.

- The level of sedation should be recorded every 15/30 minutes for the first 2 hours and then 4 hourly intervals unless clinically indicated. If the patient is sedated review 2 hourly and seek medical attention.
- If the PCA bolus/rate or lockout times are increased or parenteral opioids are increased the above observations should be repeated.
- If the patient becomes unresponsive or if the sedation score is 3 or 4 (use scoring on early warning score), then disconnect the handset, stop the PCA and contact the doctor immediately. (Follow respiratory function guidelines)

### **7.3 Nausea and Vomiting**

There are common side-effects of parenteral opioids which can be spontaneous but often brought on by movement. The presence of nausea should be assessed 4 hourly. Ensure suitable anti-emetics are prescribed and given regularly to the patient. If problems persists contact the medical team to review and change the anti-emetic regimen.

### **7.4 Pruritus**

This is a common side-effect with opioids which can manifest as localised itching or generalised itching. If this occurs prescribe Hydroxyzine 25-50mg TDS orally or other locally used anti-histamines. If pruritus persists consider changing to an alternative opioid i.e. oxycodone or fentanyl.

### **7.5 Constipation**

Reduced bowel motility is a complication of opioid therapy and requires daily monitoring of bowel function with documentation. Encourage the patient to modify their diet, increase fluid intake and the medical team to prescribe laxatives regularly.

## **8. Trouble-Shooting (according to local PCA policy)**

8.1 Changing a PCA infusion bag

8.2 Clearing the air in the PCA line

8.3 Changing the batteries

8.4 Clearing occlusions and call for service alarm

8.5 Stopping the PCA pump

## **9. Stopping the PCA or sut/cut opioids**

Patients can be weaned off once they have met following criteria:

- The patient has a pain score of none or mild pain
- The patient is able to tolerate oral medication

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- The patient has reduced the amount of PCA boluses or parenteral opioids in the previous 24 hours.
  - a) If the patient fulfils the above criteria they can be prescribed suitable oral analgesia and then stop the PCA or parenteral opioids. **Do not step down on to sub/cut opioids if stopping a PCA.**
  - b) Dose reduction should, whenever possible, be implemented in the morning and the patient's pain closely assessed. For example stop the continuous infusion first then wean the bolus dose until stopping.
  - c) Upon discharge please make sure the patient has an adequate supply of analgesia and a follow up outpatient appointment.

### **10. Contact details (according to local policy)**

If there are any medical concerns about the sickle cell patient using the PCA/Parenteral opiates contact the following: **To be completed by local sites.**

### 11. References

1. Gonzales, E.R., Bahal, N., Hansen, L.A., Ware, D., Bull, D.S., Ornato, J.P., Lehman, M.E. (1991), Intermittent Injections versus Patient controlled Analgesia for sickle cell crisis pain: Comparison in Patients in the emergency department. Arch Intern Medical July: 151 (7): 1373-1378
2. Johnson, L. (2003) Sickle cell disease patients and patient controlled analgesia. British Journal of Nursing Feb 13-26: 12 (3): 144-53
3. Rees, D.C., Olujohunfibe, A.D., Norman, E., Stephens, A.D., Telfer, P., Wright, J. (2003) Guidelines for the management of the acute painful crisis in sickle cell disease British journal of Haematology 120 (5): 744-752
4. Johnson, L. (2005) Managing acute and chronic pain in sickle cell disease. Nursing Times 101 (08): 40-44
5. Okpala, I. and Taweli, A. (2002) Management of pain in sickle cell disease. Journal of the royal college of medicine 95:456-458
6. Hines, S. and Pleasance, S. (2009) Compatibility of an injectable high strength oxycodone formulation with typical diluents, syringes, tubing's, infusion bags and drugs for potential co-administration. European Association of Hospital pharmacists 15: 32-38
7. Ogundipe, O., Person, M.W., Slater, N.G., Adepegba, T., and Westerdale, N. (1999) Sickle cell disease and nitrous oxide induced neuropathy. Clinical and laboratory haematology 21:409-412
8. National Institute for Health and Clinical Excellence (NICE) 2012. Sickle cell acute painful episode: Management of an acute painful sickle cell episode in hospital. NICE (NHS) London
9. [Sickle cell acute painful episode: management of an acute ... - NICE](#)  
[www.nice.org.uk/guidance/cg143](http://www.nice.org.uk/guidance/cg143)
10. [Sickle cell acute painful episode overview - NICE Pathways](#)  
[pathways.nice.org.uk/pathways/sickle-cell-acute-painful-episode](http://pathways.nice.org.uk/pathways/sickle-cell-acute-painful-episode)

### **Appendix 1: Nursing training (according to local policy)**

Qualified nursing staff pre-requisites for managing a patient on parenteral opioids and PCA:

- a) Registered nurses require:
  - Intravenous medication administration competence
  - Training in pain management and care of sickle cell patients as per local policy (e.g attended the Trust pain study day and sickle cell study sessions).
  - Completion of a period of supervised practice and assessed as competent in the care of patients receiving parenteral opioids and PCA.
- b) The following staff may be authorized to prepare and commence a PCA and to change the size of the bolus, lockout and drug reserve and administer a bolus:
  - Clinical nurse specialist for Haemoglobinopathies (Adult)
  - Registered nurses at band 6 and 7 and experienced band 5 nurses
  - Duty managers
  - Clinical Nurse Specialist in the pain team
- c) Registered nurses may change the prescribed PCA bag, clear air in the line, change the batteries and discontinue a PCA.
- d) Student nurses should be encouraged to observe and learn about PCA but cannot participate in the above procedures.

### **Training**

Managers have a responsibility for ensuring that as many qualified nurses as possible attend training sessions. The standard is for 75% of qualified nurses in a clinical area to have the training for PCA according to local policy. Applicants must have the support of their line manager and clinical nurse specialist in Haemoglobinopathies (Adult) or Clinical nurse specialist for pain. Advanced training to programme PCA pumps is arranged with the clinical nurse specialist for haemoglobinopathies (adult) or Clinical Nurse Specialist for pain. All nurses must fulfil all competencies for PCA before being independent with this practice (See appendix three). Reassessment of competencies

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must be performed annually to ensure that the nurse remains competent and confident to deliver care.

### Appendix 2: Assessor's competency checklist for nurses administering opioids via patient controlled analysis (PCA)

Name of Nurse

Name of Assessor

Competences	Evidence	Criteria met Yes/No
Knowledge of physiology of pain	Demonstrate an understanding of the physiology of pain	
Can articulate the criteria for choosing PCA: Indications/contraindications	Understanding indications and contraindications of PCA	
PCA Regime	Able to explain correct Diamorphine/Morphine/Oxycodone/Fentanyl regime differences	
Explain application of pump to the sickle cell disease patient	Can explain use of PCA in relation of pain control	
Explain side-effects of Diamorphine/morphine/Oxycodone/Fentanyl	Know how to act on side-effects Able to discuss effects and identify side-effects of medication	
PCA Policy Emergencies	Understand the policy and who to contact in an emergency  Verbalise the clinical incident reporting	

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	<p>policy</p> <p>Understands accountability in relation to PCA management</p>	
Pump settings	<p>Understanding the following terms:</p> <ul style="list-style-type: none"> <li>• Bolus dose</li> <li>• Lockout</li> <li>• Continuous dose/rate</li> </ul>	
Preparation and commencing PCA	<p>Refers to PCA prescription</p> <p>Performs correct procedure for priming the infusion line</p>	
Programming the PCA	<p>Demonstrate how to programme diamorphine/morphine/ Oxycodone regime</p> <p>Programme the PCA according to prescription</p> <p>Locking the PCA</p> <p>Labelling the line</p>	
Managing the PCA pump	<p>Can clear the air in the line</p> <p>Can change the PCA bag</p> <p>Can change the batteries in the pump</p> <p>Can check the programme</p>	
Nursing care	<p>Can give rationale and frequency of</p>	

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	<p>monitoring:</p> <ul style="list-style-type: none"> <li>• Pain score</li> <li>• Sedation score</li> <li>• Nausea score</li> <li>• Respiratory rate</li> <li>• Oxygen saturations</li> <li>• Check sub/cut site</li> <li>• Appropriate action to be taken should complications develop</li> <li>• Documentation</li> </ul>	
<p>Stopping PCA</p> <ul style="list-style-type: none"> <li>•</li> </ul>	<p>Able to tell:</p> <ul style="list-style-type: none"> <li>• The weaning criteria                             <ul style="list-style-type: none"> <li>• Step down analgesia</li> </ul> </li> </ul>	

Nurse's signature

Assessor's signature

Date