University Hospitals of Derby and Burton NHS Foundation Trust

# Management of an Acute Painful Crisis in Sickle Cell Disease - UHDB Clinical Guideline

**Reference No: CG-T/2013/170**

**Adults with Sickle Cell Disease presenting with an acute painful sickle cell episode should be treated as an acute medical emergency.**

Sickle cell disease affects up to 15000 people in the UK (ref). Many of these patients will present acutely to a variety of hospitals in the country and may not be known already to staff at the hospital.

Sickle cell disease (either HbSS or compound heterozygote states Hb SC, HbSβ Thal or other compound states) commonly gives rise to acute painful crises. The pain may occur in any part of the body but most commonly is in the limbs. Pain in the chest or abdomen should prompt consideration of alternative causes of pain in those sites (eg MI, acute appendicitis). Sickle pain is very severe usually requiring opiate analgesia and may have a pain score of up to 10/10. It is usually clear to the patient that this is their 'usual' sickle pain (i.e patients with sickle cell disease are the experts in their own painful crises).

The pathogenesis of sickle cell pain is thought to be due to deoxygenated sickle Hb forming large polymers which cause red cells to become deformed and show irreversible sickling leading to small vessel obstruction. Triggering factors include cold, dehydration and infection but often no specific trigger is found.

Patients with sickle cell disease often have a chronic anaemia of 50-100g/L which may be normal for them. Blood transfusion is **NOT** routinely indicated and in fact may exacerbate a crisis. **Blood transfusions should only be used after advice from the Haematologist on call.**

**Common Presentations include:**

· severe pain

· pyrexia, tachycardia, tachypnoea, hypotension

· increased pallor, breathlessness, exhaustion

· chest pain, consolidation on CXR, $SaO_2$ <94% on air

· abdominal pain or distension, diarrhoea, vomiting

· severe thoracic/back pain

· headache, drowsiness, CVA,TIA or any abnormal CNS signs

**Arrangements for Admission at RDH (adults)**

During working hours, patients are advised to contact the Haematology Clinical Nurse Specialist on their mobile via the switchboard at Royal Derby Hospital or the on call Haematologist out of hours. The patient will be told where they can be seen ie CTAU, a Haematology ward or Medical Assessment Unit (MAU). Patients via ambulance, self-presentation and some patients not known to the service may be admitted to A+E /MAU without knowledge of the Haematology team. In all cases the on-call Haematologist must be informed of the admission by the Haematology CNS during working hours and by A&E/MAU during out of hours.

If seen on MAU and requiring admission for longer than 18 hours, patients will normally have to be moved to another ward: ideally ward 301 if bed available. (Sickle cell disease is a multi-system disorder and referral to non-haematology specialists may be advised but the haematology team will remain involved.)

**Assessment and Initial Management of Patient with Sickle Cell Disease**

Overall the plan for these patients is an urgent assessment with review of their pain and observations immediately. Generally, patients will require admission as the pain is uncontrolled with their normal home pain management. Baseline observations must be performed as standard and NEWS documented. Escalation should be carried out in line with RDH policy if the NEWS score warrants this. If opiates have been administered by the ambulance crew, this MUST be documented clearly. Baseline oxygen saturations must be documented ON AIR. If <95% then an ABG should be performed and more urgent escalation to haematology considered. Suspected sepsis should be managed in line with RDH sepsis policy.

**PAIN:** If the pain is typical sickle cell pain uncontrolled with home analgesics and observations are stable and there are no unusual features then analgesia should be given as below. **Analgesia must be administered within 30 minutes of admission (NICE CA143).**

Check previous drug charts and patients care plan for reference AND

Give a combination of analgesics for adequate and quicker pain control. escalate rapidly with severe pain.

Regular **Paracetamol** 1g Four times a day - IV if necessary then Oral.

PLUS

Regular **Ibuprofen** 400mg Three times a day (if not contraindicated).

PLUS

**Morphine** by the most appropriate route. Oral should not be offered as it is ineffective for acute severe pain. Subcutaenous is the most appropriate route in this

setting, unless the patients present to ED in crisis, where it is initially appropriate to titrate IV morphine to effect, before moving on to SC for maintenance. Some patients may prefer oral morphine, but if in acute severe pain, should be offered IV or SC initially.

If the patient does not have a dedicated analgesia plan, then doses are based on weight

If < 50Kg give 5mg morphine sulphate SC stat

If > 50Kg given 10mg morphine sulphate SC stat

If IV, give up to 20mg titrated to effect.

**Efficacy of analgesia should be assessed 30 minutes after IV morphine (or 1 hr after SC)** and a further dose of morphine administered provided that there is no evidence of opioid toxicity.

If the RR is < 10 per minutes, omit opioid analgesia

If RR < 6 give Naloxone 100micrograms every 2 minutes as necessary

**Pain should then be reassessed every 30 minutes until adequate pain control is achieved**.

If pain control is inadequate despite 2 consecutive doses of Morphine

Discuss the case with the oncall haematology registrar or the CNS (during working hours)

Consideration of escalation to **patient controlled analgesia** (see below)

Consideration of alternative causes of pain if out of context with 'usual' sickle pain

Once pain is controlled, regular adjunct analgesia and PRN SC opiate analgesia (0.05 - 0.1mcg/Kg to nearest mg PRN as per protocol) should be prescribed. Please see full SC OPIOID protocol on KOHA. Summary attached

**Adequate analgesia should be achieved by 60 minutes. This will reduce the requirement for morphine and avoid the need for PCA.**

Once pain is controlled, observations must be done HOURLY for first 6 hrs and then 4hourly.

**Inform the Acute Pain Team Nurses early via bleep if the sickle pain is not controlled with SC or IV morphine by 60mins and they will review and advise appropriately. PCA is currently available on Ward 301 and is being looked into on MAU. Please see separate PCA guideline on KOHA.**

**For persistent pain after 90min contact the ITU outreach team/ITU on-call for**

**consideration for a PCA after discussion with the on-call Haematologist.**

Laxatives, antiemetics and anti-pyretics should be prescribed usually on a PRN basis

Alternative analgesics to consider Oxycodone, Entonox (only when available and staff trained to use) NB, PCA is for standard morphine only, no other opiates/regimes are available via PCA outside of critical care.

**Alteration of dose of analgesia**

Requirements for analgesia need to be reconsidered on a daily basis. When the pain is well controlled aim to reduce dose by small amounts and switch to oral equivalents when patient is clearly improving (pain score /chart may be helpful)

## Investigations

Patients presenting with sickle cell crisis require the following investigations

· FBC and reticulocyte count

· Group and save

· U&E, LFT, LDH, CRP (LFTs may be obstructive if gallstones)

· CXR

· Blood cultures, MSU, Throat swabs for serology

· ABG if < 95% on air

· XR is indicated if a patient develops localised bone pain and fever as osteomyelitis is a recognised and potentially serious complication of sickle cell disease.

· COVID PCR

· Hb electrophoresis for a new/unknown patient

· If transfused within 12 weeks, DAT

# Pain management Overview

| Time from 1st review | Reason | Action | Comments |
|---|---|---|---|
| 0-90 mins | Immediate control acute pain<br><br>Supportive measures | 1. Paracetamol can be given IV in acute phase<br>2. Ibuprofen if no contraindication to NSAIDs<br>3. Morphine 0.15 mg/kg IV/SC and titrate until pain controlled and commence frequent observations every 5 minutes for 30 minutes<br>4. Reassess pain and side-effects of morphine regularly<br>5. Prochlorperazine or cyclizine, chlorpheniramine, lactulose + senna - should be prescribed as PRN<br>6. Enoxaparin 40mg sub-cutaneous for thromboprophylaxis<br>7. Intravenous fluids: 2-3 litres over 24hours<br>8. Oxygen – maintain O2 saturations >95% at all times | **All these action measures are essential upfront** |
| >90 mins<br><br>**OR** | Continued Intermittent given by staff | Morphine 0.15 mg/kg SC PRN as per protocol. (Please see SC Opioid protocol on KOHA)<br><br>Observations at least HOURLY for the first 6 hours | |
| >90mins | PCA (patient controlled analgesia) | Follow the Trust guidelines on PCA MORPHINE ONLY if pain control is inadequate at 90 minutes<br><br>Monitor the pain<br><br>Record the pain score and sedation score on the Acute Pain Chart | **Avoid delays in starting PCA** |
| Every 30 mins then hrly | Monitor | Pain, sedation, vital signs, respiratory rate until pain controlled and stable then every 1 hour | |

## Sedation assessment

| **Patient** | **Sedation score** | **Action** |
|---|---|---|
| Awake and alert | 0 | Continue with regular observations |
| Drowsy, easy to rouse | 1 | If respiratory rate <10/min, give 40% oxygen + inform the ITU outreach team/ on-call ITU doctor |
| Very drowsy, difficult to rouse | 2 | If respiratory rate <6/min, give 100% Oxygen and Naloxone. Hold opioids. Inform the outreach team. |

| | | |
|---|---|---|
| Normal sleep, easy to rouse | S | Continue with regular observations |

Can I add the summary s/c morphine/pethidine algorithm here- suggestion of the acute pain team

**Oxygen**

- There is no good evidence for this being used routinely in all cases of painful crisis
- Use should be dictated by the clinical situation and oxygen saturations:
- If $SaO_2$ <95% on air, give $O_2$ by face mask
- Check arterial gases if $SaO_2$ on air is <95%
- Monitor $SaO_2$ while patient is on supplementary oxygen aiming to keep $O_2$ level > 98%
- If arterial $pO_2$ ($PaO_2$) <10.7kPa use mask giving 35% inspired oxygen
- If arterial $PaO_2$ < 9.3kPa seek additional help – generally by involving HDU and outreach team

**NOTE if there is an increasing O2 requirement urgent medical review is required to exclude acute chest syndrome**

**Thromboprophylaxis**

Sickle cell disorders are associated with an increased thrombotic potential

All patients should have a formal VTE assessment

All patients who are admitted with a severe sickling crisis who are immobile should be commenced on low molecular weight heparin prophylaxis, unless contraindicated.

Do NOT use TED stockings

**Antibiotics**

If the patient is febrile or has a history suggestive of an infective cause of the crisis they should be commenced on antibiotics

Commence antibiotics in line with RDH microbiology guidance.

Co-amoxiclav is the antibiotic of choice in most cases unless penicillin allergic.

Most patients are on oral penicillin maintenance which can be stopped and restarted when the course of alternative antibiotics has been completed.

**Transfusions**

Patients with sickle cell disease often have a chronic anaemia of 50-100 g/dl

Hb S has a lower oxygen affinity than Hb A so oxygenation is better than expected

It is useful to check the patient's steady state Hb level when reviewing their blood

count.

The Hb may fall 1-2 g during a sickle crisis but blood transfusion is **NOT** routinely indicated and in fact may exacerbate a crisis

Folic acid - all sickle cell patients should be on this regularly at 5mg od

**Blood transfusions should only be used after advised by the Haematologist**

Consider if there are symptoms of severe anaemia or if the Hb has fallen > 20g and is below 50 g/dl.

Check HbS% before transfusion.

The transfusion should aim to return the Hb to the steady state level and the **blood should be fully phenotypically matched.**

**Exchange Transfusion**

The aim of an exchange transfusion is to replace the sickle blood with non sickle and needs to be discussed with the on call Haematologist.

The indications for urgent exchange transfusion are for

Sickle chest crisis – see separate guidance

Cerebral sickling

Multiorgan failure

·Red cell exchange is preferably done using the cell separator via the stem cell nurses at Haematology Day Case Unit, Nottingham City Hospital. It is a consultant to consultant referral. Manual exchange transfusion may be necessary and performed in an emergency

· Routine elective exchange transfusion may be done for recurrent severe crises and pre-operatively

· Blood should be fully phenotyped for the patient and Blood Bank should be contacted as soon as an exchange transfusion is planned

· Hb A and S levels should be sent to the Haematology laboratory before and after manual exchange transfusion.

· A Hb S level of < 30% and Hb of 90g/dl is the goal of this therapy

**Monitoring Compliance**

| What will be measured to monitor compliance | How will compliance be monitored | Monitoring Lead | Frequency | Reporting Arrangements |
|---|---|---|---|---|
| NICE pain score audit data | Regularly submitted to NHS England haemoglobinopathy dashboard and reviewed at East Midlands haemoglobinopathy network | Caroline Harvey | Annually | Via Dashboard |

## References

Guidelines for the management of the acute painful crisis in sickle cell disease. Rees et al, British Journal of Haematology 2003, 120: 744-752.

Sickle cell acute painful episode: management of an acute painful sickle cell episode in hospital, NICE clinical guideline 143, June 2012

Adopted from East Midlands Network Guideline, Nottingham City Hospital.

Trust guidelines on IV and Sub-cutaneous morphine; and PCA, Royal Derby Hospital.

## Documentation Controls

| | |
|---|---|
| **Development of Guideline:** | **Consultant Haematologist** |
| **Approved By:** | **CDCS - Dec 2023** |
| **Date of Approval:** | **Dec 2023** |
| **Review Date:** | **Dec 2026** |
| **Key Contact:** | **Consultant Haematologist** |