Sickle Cell Disease (SCD)
Acute pain crisis: a medical emergency
Acute pain crisis = Vaso-occlusive crisis (VOC)

An acute presentation of pain in patients with SCD may represent a VOC, an acute complication of SCD, exacerbations of chronic complications of SCD, or SCD-unrelated conditions. This document highlights the importance of assessing and managing sickle cell pain alongside standard clinical assessment.

Acute presentation

Observations
T, P, BP, RR, weight, sats on air
Sats 3% below norm or ≤ 95% is high risk for acute chest syndrome
If febrile: commence local sepsis protocol as patients hyposplenic

Immediate pain assessment
Document:
- pain score 0-3
- analgesia taken pre-arrival & usual regime
- location/duration/description
- triggering factor
Ask the patient typical SCD pain

Clinical assessment to determine cause of pain

Likely VOC

Other causes of pain
- Follow condition-specific pathway
- Ask about recent transfusion? delayed transfusion reaction presenting with pain

Investigations
EPR careset (listed overpage)
CXR if signs/symptoms/low sats

Interventions & Management plan
- Fluids (3-4L/day), fluid balance
- Abx if pyrexial
- Incentive spirometry
- VTE assessment
- Document a clear pain management plan

Reassess every 30 min
- Repeat pain score
- Escalate as per analgesic titration
- Optimise multimodal analgesia

Notify Haematology of patient presentation

Repeat regular observations as clinically indicated – at least 4-hourly
Blood tests

<table>
<thead>
<tr>
<th>Test</th>
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<tbody>
<tr>
<td>FBC</td>
<td>CRP</td>
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<tr>
<td>Retic count</td>
<td>Clotting screen</td>
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<td>Iron studies</td>
<td>MRSA screen</td>
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<td>U&amp;Es</td>
<td>Blood cultures</td>
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<tr>
<td>LFTs &amp; GGT</td>
<td>Urine cultures</td>
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<tr>
<td>Ca, Mg, PO4</td>
<td>G&amp;S</td>
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<tr>
<td>LDH, glucose</td>
<td>Hb HPLC (haemoglobinopathy screen)</td>
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Pregnant women

Analgesia as per this protocol unless specific plan in place. Opiates are acceptable. No NSAIDs 2nd trimester.

Notify obstetric team of admission.

SCD-related complications

Acute chest syndrome
- Fever
- Chest pain, SOB
- Hypoxia
- Abnormal respiratory signs/symptoms
  - If sats <95% or 3% below normal baseline → ABG
  - CXR

Protocol here

Acute neurological presentation
- Early involvement of haematology and neurology teams
- Early G+S sample for possible exchange transfusion
- Rehydrate immediately

Protocol here

Acute abdominal presentation
- Cholecystitis is a common complication of SCD

Protocol here

Sepsis
- High risk as all SCD patients are functionally hyposplenic
  - Consider pneumococcal sepsis, gram negative sepsis, osteomyelitis
  - Consider neutropenia if patients on hydroxyurea
  - Instigate local sepsis protocol

SCD sepsis Protocol here

Aplastic crisis
- Acute drop in Hb
- Low retic count
- Check parvovirus B19 IgM and PCR
- May need transfusion
- Involve haematology

Protocol here

Priapism
- A sustained painful erection
  → it may be necessary to elicit this by direct questioning
  This is a urological emergency
  - Involve haematology and urology teams
  - Pain management & fluids

Protocol here

Red Cell Transfusions
- Top-up and/or exchange transfusions form part of the management of some of these complications
  - Involve haematology

Protocol here

Other protocols available at:
http://nssg.oxford-haematology.org.uk/red-cell/red-cell.html
General principles

The haematology team should be made aware of ALL admissions. Patients should be looked after on a haematology ward whenever possible.

Do not cannulate lower limbs due to high risk of thrombosis and ulceration.

When prescribing analgesia, assess what the patient has already taken prior to presentation, also what they normally take/require.

Subcutaneous opioids should be prescribed initially for immediate pain relief as cannulation may result in a delay in opioid administration. IV PCA is best but SC PCA can be prescribed.

Pain Assessment

- Ask about individual coping strategies

- The OUH Trust pain assessment scale is 0 = no pain; 1 = mild pain; 2 = moderate pain; 3 = severe pain

- Assess pain descriptors, location, triggers and levels of anxiety in addition to intensity.

Analgesia titration

Mild (0-1): paracetamol +/- NSAIDs if appropriate*

Moderate (2): add 30-60mg codeine phosphate regularly 6-hourly OR tramadol 50-100mg 6-hourly OR dihydrocodeine 30-60mg 6 hourly (max 240mg in 24hrs) WITH oral morphine IR 10mg 4-hourly OR oxycodone 5mg IR 4-hourly (increase to 2-hourly if needed for breakthrough pain).

Severe (3): assess for PCA (preferably IV morphine-oxycodone if morphine contra-indicated). Only commence background infusion after discussion with pain service. Continue all other adjuvant analgesics.

If patients are on long term MST or OxyContin (etc.) - please refer to pain service for advice, or contact on-call anaesthetist out-of-hours.

Refer to local guidelines for PCA prescription and monitoring

Key principles:

- Prescribe: Anti-emetics, anti-pruritics, laxatives and naloxone
- Location of patient in a ward where PCA use has been agreed
- Observations must be carried out regularly (e.g. every 30 min until pain is settled then 2 hourly)
- Ensure PPI with NSAID- avoid diclofenac. First line use Ibuprofen
- Do not use pethidine
- Refer to Pain Service for input.

Specific contraindications to recommended pharmacological agents may apply in individual patients (e.g. *pregnancy, *renal dysfunction, epilepsy, concurrent TA/SSRIs if using Tramadol, etc.)
**App guidance**

The Oxford PainGuide is now available on the MicroGuide app platform. The guidelines have been written by the OUHFT Pain Service in collaboration with pharmacy and specialist medical teams to provide evidence-based, user-friendly and up to date recommendations. Simply download the MicroGuide app from the App Store, go to “get Guides” and select OUH, then select “Pain Guidelines”.

**Discharge planning and specialist review**

- Prescribe reducing dose of analgesia (max 3 days opiates)
- Notify CNS for 48-72 hr post discharge review
- Plan 2 - 6 week review in clinic depending on presentation
- Refer to Red Cell MDT if new or complex issues have arisen

**Audit**: Acute SCD pain management is subject to annual audit as per NICE guidelines

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**Review**

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