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

**Likely VOC**

**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

**Clinical assessment to determine cause of pain**

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

**Immediate pain assessment**
REfer to patient pain plan

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

**Analgesia within 30 min of presentation**
- Based on prior use, pain score and analgesic titration unless patient has handheld record or EPR entry

**Reassess every 30 min**
- Repeat pain score
- Escalate as per analgesic titration
- Optimise multimodal analgesia
- Continue until satisfactory pain relief has been achieved

**Notify Haematology of patient presentation**

**Repeat regular observations as clinically indicated – at least 4-hourly**
**Acute chest syndrome**
- Fever
- Chest pain, SOB
- Hypoxia
- Abnormal respiratory signs/symptoms
  - If sats <95% or 3% below normal baseline \(\rightarrow\) ABG
  - CXR

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

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

**Acute abdominal presentation**
- Cholecystitis is a common complication of SCD
- Splenic/hepatic sequestration are rare but serious complications
- Add amylase if abdo pain

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

**SCD-related complications**

**Blood tests**
- FBC
- Retic count
- Iron studies
- U&Es
- LFTs & GGT
- Ca, Mg, PO4
- LDH, glucose
- CRP
- Clotting screen
- MRSA screen
- G&S
- Urine cultures
- Blood cultures

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

**Pregnant and Breastfeeding: women**
Analgesia as per this protocol unless specific plan in place.
Opiates are acceptable.
NSAIDs: use with caution before 12/40. Avoid after 31/40.
SCD Obstetric 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.

Severe (3): assess for PCA (1st line IV morphine-oxycodeone if morphine contra-indicated). Only commence background infusion on advice of Pain service. Continue all other adjuvant analgesics.

If patients are on long term opioids such as MST or oxycodone MR - please refer to pain service for advice, or contact on-call anaesthetist out-of-hours.

Specific contraindications to recommended pharmacological agents may apply in individual patients (e.g. *pregnancy, *renal dysfunction, epilepsy).

Refer to local guidelines for PCA prescription and monitoring

Key principles:

• Prescribe: Anti-emetics, anti-histamine, laxatives and naloxone

• Location of patient in a ward where PCA use has been agreed.

• Observations must be carried out regularly (e.g. every 15 mins for first hour and 30 min for next 2 hours until pain is settled. Then 4 hourly whilst PCA in situ.)

• Ensure PPI with NSAID- avoid diclofenac. First line use Ibuprofen

• Do not use pethidine

• Refer to Pain service for advice if analgesic regimen fully optimised and pain remains severe.

Please contact the Pain Service for advice as needed.
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 opioids)
- 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

Authors: Rewrite October 2020
Oxford University Hospitals (OUH) Haematology: Sandy Hayes, Dr Noemi Roy
OUH Pain Service: Dr Jane Quinlan (Consultant Anaesthetist), Felicity Wintle (Advanced Clinical Practitioner)

Review

<table>
<thead>
<tr>
<th>Name</th>
<th>Revision</th>
<th>Date</th>
<th>Version</th>
<th>Review date</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wale Atoyebi, Deborah Hay</td>
<td>Pre-peer review</td>
<td>Jan 2013</td>
<td>1.1</td>
<td>Jan 2015</td>
</tr>
<tr>
<td>Deborah Hay</td>
<td>Routine review</td>
<td>Aug 2015</td>
<td>1.2</td>
<td>Jan 2017</td>
</tr>
<tr>
<td>Deborah Hay</td>
<td>ODN meeting</td>
<td>Oct 2016</td>
<td>1.3</td>
<td>Oct 2018</td>
</tr>
<tr>
<td>Rewrite authors as above</td>
<td>Rewrite</td>
<td>Nov 2020</td>
<td>2.0</td>
<td>Nov 2022</td>
</tr>
<tr>
<td>Felicity Wintle, Jane Quinlan, Wale Atoyebi</td>
<td>Full Review</td>
<td>May 2024</td>
<td>3.0</td>
<td>May 2026</td>
</tr>
</tbody>
</table>