Orthopaedic complications in patients with Sickle Cell Disease (SCD)

Avascular Necrosis (AVN)
This complication may start in adolescence and often gives rise to chronic pain and limitation of movement due to joint damage, rather than on-going vaso-occlusion.

Presentation
- Pain in the hip, groin, knee or shoulder on movement; later at rest. Repeated or prolonged pain (>8 weeks) should be investigated for avascular necrosis.
- Limitation of movement; particularly abduction and external rotation of the hip, external rotation of the shoulder.
- Differential Diagnosis:
  - Osteomyelitis and or septic arthritis, which may present together and are associated with swinging pyrexia, severe systemic disorder, positive blood cultures, high CRP/white cell count and pain

Investigations
- Plain X-ray, if required by Radiology
- MRI: will show changes much earlier than x-ray

Management
- Paracetamol plus NSAIDs with H2 antagonists, or codeine derivatives.
- Rest and the avoidance of weight bearing (very difficult to implement).
- Transfusion cannot reverse the process but may prevent progression in the contralateral joint
- Refer for orthopaedic assessment and treatment:
  - Osteotomy and/or decompression surgery may be considered.
  - Major joint surgery may be necessary if pain is continuous (>2 years) or very severe, or if the patient’s mobility is seriously affected.
  - Different types of prosthesis, hip fusion, or bone grafting are used depending on the individual case. Infection is not uncommon.
  - The possibility of failure, the likelihood of some residual pain, the potential life of the prosthesis, and the limitations imposed must always be discussed with the patient pre-operatively by the surgical team.

Orthopaedic surgery for AVN Thames Valley Pathway
- Referrals to OUH (Oxford) NOC Orthopaedic Service are encouraged due to their experience in managing these complex cases. There is also acute haematology/red cell exchange support if required.

- A Haematology exemption form should be completed and sent with the referral
Wessex and Thames Valley Haemoglobinopathy Network

Adult Haemoglobinopathy Service

- Surgical and anaesthetic teams must be forwarded a copy of the guidelines for pre/perioperative management and transfusion of patients with sickle cell disease. These protocols should also be added to the patient electronic record.

Perioperative management of patients with Sickle Cell Disease

Transfusion prior to elective surgery in patients with Sickle Cell Disease

- Blood bank must be alerted in advance to ensure that appropriate extended phenotyping of blood is undertaken. Any communication with blood bank must include sickle cell disease as a diagnosis

- Exchange transfusion will be required for all significant orthopaedic surgery.

OUH NOC Orthopaedic surgery referral contacts:

<table>
<thead>
<tr>
<th>Orthopaedic - ankle</th>
<th>Mr Bob Sharp, Consultant orthopaedic surgeon, Nuffield Orthopaedic Centre (NOC)</th>
<th><a href="mailto:Footandankle.noc@nhs.net">Footandankle.noc@nhs.net</a></th>
</tr>
</thead>
<tbody>
<tr>
<td>Orthopaedic - hip</td>
<td>Mr McLardy-Smith and Mr Simon Newman, Consultant orthopaedic surgeons, NOC</td>
<td><a href="mailto:Hipandknee.noc@nhs.net">Hipandknee.noc@nhs.net</a></td>
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<tr>
<td>Orthopaedic - reconstruction</td>
<td>Mr Martin Mcnally, Consultant orthopaedic surgeon, NOC</td>
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<tr>
<td>Orthopaedic – shoulder and hand</td>
<td>Mr Chris Little, Consultant orthopaedic surgeon, NOC</td>
<td><a href="mailto:Hand.noc@nhs.net">Hand.noc@nhs.net</a></td>
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Wessex Pathway
The Wessex Network is in process of developing their own pathways.

Osteomyelitis and Septic Arthritis
Osteomyelitis and septic arthritis are serious and potentially disabling conditions but can be difficult to differentiate from benign conditions of SCD, such as vaso-occlusive crisis. Diagnosis requires careful evaluation of the clinical presentation and history, laboratory testing, and imaging. There could be multiple sites of presentation and recurrence many years later due to bacteria forming a long-lasting biofilm. They are most commonly caused by salmonella species, staphylococcus aureus and gram-negative enteric bacteria.

Treatment of osteomyelitis in patients with SCD may be medical or surgical, but considerations in antibiotic selection and management preoperatively and postoperatively must be taken to ensure optimal outcomes.

Septic arthritis treatment involves IV antibiotics and may also require arthroscopic surgical washout.

Presentation
Patients may present with fever and a warm painful, swollen limb or area over the bone, with impact on mobility. Usually, osteomyelitis affects the diaphysis of long bones, such as the femur and humerus. Septic arthritis can involve any joint.
Diagnosis and Management
- Detailed history, particularly of childhood admissions (if possible) as recurrent osteomyelitis is not uncommon
- Blood cultures
- Ultrasound and/or MRI.
- Refer to the national bone infection unit at NOC (boneinfection.noc@nhs.net) or local team
- Microbiology advice
- Consider midline or PICC line insertion early in the course of antibiotic therapy
- Pain relief, IV fluids and appropriate antibiotics

Referrals from other HCC/ SHT teams
Referred Sickle Cell Patients, from other networks/ specialist haemoglobinopathy teams to the OUH NOC are followed up by the Oxford Red Cell Clinical Team.

References
Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK © Sickle Cell Society 2018


Management of Osteomyelitis in Sickle Cell Disease: Review Article
Al Farii, Humaid MD, MRCS (Ir); Zhou, Sarah MD; Albers, Anthony MDCM, FRCSC, JAAOS: Global Research and Reviews: September 2020 - Volume 4 - Issue 9 - p e20.00002-10. doi: 10.5435/JAAOSGlobal-D-20-00002

Authors:
Dr Wale Atoyebi, Clinical Lead for Adult Haemoglobinopathies

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<tr>
<td>Dr Wale Atoyebi</td>
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<td>Dr Deborah Hay</td>
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