NHS West London Haemoglobinopathy Coordinating Centre
A partnership between
NHS Imperial College Healthcare NHS Trust | NHS London North West University Healthcare NHS Trust | NHS St George's University Hospitals NHS Foundation Trust

GP PRIMARY CARE GUIDE: SICKLE CELL DISEASE

What is Sickle Cell Disease?

- Inherited haemoglobinopathy caused by the sickle mutation (HbS).
- HbSS = sickle cell anaemia; HbSC and sickle-β-thalassaemia also clinically significant.
- Sickling occurs when deoxygenated HbS polymerises → vaso-occlusion + haemolysis.
- Prevalent in African, Caribbean, Mediterranean, Middle Eastern, Indian populations, effecting ~18k in the UK

Red Flags Requiring Urgent Hospital Referral

- Severe bone pain needing opiates
- Pallor, breathlessness, exhaustion
- Fever >38°C, tachycardia, tachypnoea, hypotension
- O_2 sats <95% on air
- Chest pain, crackles, suspected ACS
- Severe abdominal pain/distension
- Neurological symptoms (weakness, seizures, altered consciousness)
- Priapism
- Suspected sequestration or aplastic crisis

Key Acute Conditions

Acute Chest Syndrome

- Chest/abdominal pain, fever, tachypnoea, cough, desaturation.
- Urgent hospital referral; give high-flow oxygen and analgesia.

Stroke

- Limb weakness, speech/visual disturbance, seizures, behavioural change.
- Painless limp in children is a red flag.
- Immediate hospital transfer.

Abdominal Pain

- Consider abdominal crisis, girdle syndrome, sequestration, appendicitis, cholecystitis, pancreatitis.
- Girdle syndrome & sequestration = emergency admission.

Aplastic Crisis (Parvovirus B19)

- Sudden severe anaemia, low reticulocytes.
- Check FBC, retics, G&S.
- Admit if symptomatic or Hb significantly low.

Splenic/Hepatic Sequestration

- Rapidly enlarging spleen/liver, shock, severe anaemia.
- Immediate hospital transfer.

Priapism

- Painful erection >2 hours = emergency.
- Stuttering priapism (<4 hours) requires early intervention.

> Safety net advice to give every SCD patient (return immediately for fever ≥38 °C, chest pain/breathlessness, new neurological deficits, priapism ≥2 h, rapidly enlarging spleen/abdomen, dark urine/jaundice post transfusion).

Management in Primary Care

Painful Vaso-Occlusive Crisis

Give analgesia within 30 minutes:
If pain is controlled and patient has a documented individual care plan, consider supported home management with very clear safety netting and rapid re access; otherwise hospital daycare or ED

1. Paracetamol ± NSAID
2. Add weak opioid (codeine)
3. Add strong opioid (oral morphine)

- Only one dose in primary care → refer to hospital.

Hydration

- Encourage oral fluids.
- During illness: aim for 150% maintenance.
- Watch for dehydration (hyposthenuria common).

Infection

- If mild/no focus: increase Penicillin V to treatment dose.
- If infection suspected: Co-amoxiclav ± macrolide (local guidelines).
- If systemically unwell → hospital.

Oxygen

- If sats <95% on air → refer. Compare with baseline (some patients have lower baseline).

> Phone the haemoglobinopathy when persistent pain despite two oral morphine doses; SpO_2 drop to ≤95%; fever/sepsis).

Diagnosis & Screening

Testing
- HPLC + confirmatory electrophoresis/solubility test.
- Offer to all people of childbearing age.

Antenatal
- Test pregnant women before 10 weeks if status unknown.
- If both parents carriers → refer for genetic counselling.

Newborn Screening
- Heel-prick test at 5–8 days.
- GP responsibilities:
- Record result in notes.
- Prescribe penicillin V from 3 months.
- Prescribe folic acid.
- Ensure full immunisation schedule.
- Encourage parental testing if needed.

Clinical Presentations

Mechanisms
- Vaso-occlusion → pain, ACS, stroke, organ infarction.
- Haemolysis → anaemia, nitric oxide depletion → pulmonary hypertension, priapism.

Acute Complications
- Painful crisis
- Acute chest syndrome (ACS)
- Infection/sepsis
- Splenic/hepatic sequestration
- Aplastic crisis
- Stroke
- Priapism
- Abdominal crises
- Osteomyelitis

Chronic Complications
- Fatigue, retinopathy, osteopenia/AVN, pulmonary hypertension, chronic lung disease, renal impairment, gallstones, leg ulcers, delayed puberty.

Chronic Care & Monitoring

Pulmonary Hypertension
- Specialist ECHO every 3 years.
- GP: monitor breathlessness, O_2 sats.

Renal Disease
- Annual urine ACR from ~age 10; if albuminuria → ACEi/ARB and specialist review; monitor eGFR/U&E regularly; avoid NSAIDs in CKD

Retinopathy
- Annual ophthalmology review. HbSC may have higher proliferative retinopathy risk, support strict annual review.

Growth & Puberty
- Monitor height/weight; delayed puberty common.

Treatment Overview

Prophylaxis
- Penicillin V from 3 months (lifelong).
- Folic acid daily.
- Vaccinations:
- Routine childhood schedule
- Pneumovax II at 2 years, then every 5 years
- Annual flu
- Hepatitis B + boosters

Hydroxycarbamide
- First-line disease-modifying therapy.
- GP: monitor for cytopenias; urgent FBC if fever or symptoms.

Transfusion Programmes
- For stroke prevention, organ damage, recurrent crises.
- GP: monitor for post-transfusion symptoms (fever, jaundice, dark urine).
- Ensure extended antigen matching recorded in notes
- Iron overload monitoring (serum ferritin, FerriScan, MRI T2*).

Adolescence, Transition & Pregnancy
- Transition to adult services requires planning.
- Pregnancy = high-risk → joint obstetric/haematology care.
- Partner testing essential.

GP Quick Actions Checklist
- Record genotype clearly.
- Ensure prophylaxis + vaccinations.
- Provide prompt analgesia.
- Maintain low threshold for referral.
- Avoid NSAIDs in renal impairment.
- Encourage hydration and infection vigilance.
- Liaise early with haemoglobinopathy team.

BMJ course for GPs:
https://new-learning.bmj.com/course/30000376

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