SICKLE CELL CRISSES
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At least 500 patients with sickle cell diseases (HbSS, HbSC, HbSBthal) live in the St George’s catchment area. Intaking teams can expect to see >100 crises/year. Many patients have a personal management protocol which is kept in a file in their name in A&E Majors. Copies of patient protocols are also on EPR under electronic documents and should be consulted for advice on prompt initial treatment, since it may differ in important details from the generic advice given below.

PAIN CRISIS
The most common type of crisis presents as agonising and relentless pain. The pain may be localised to a single long bone, present symmetrically in several limbs, or involve the axial skeleton (lumbar spine, ribs or pelvis). Pain can lead to behavioural changes including becoming non-communicative or occasionally panicked and aggressive. If pain is bad enough to bring the patient to hospital, the patient usually warrants admission. Patients will often have tried a variety of analgesics at home including some form of opiate.

In the Accident and Emergency Department there are ED guidelines available.

Achieving fast and adequate pain control is the priority.

- Patients with sickle cell disease should be triaged as urgent.
- Nurse assessment with vital sign observations
- If pain crisis: administer analgesia as per protocol if one is available, or as per ED guidelines
- Analgesia must be given within 30 mins
- Assess pain every 30 mins until adequate pain relief has been achieved
- For parenteral analgesia the subcutaneous route is preferred to intramuscular (to preserve muscles)
- If no protocol and requiring parenteral opiate then 0.1mg/kg morphine sc is an appropriate starting dose. This can be repeated at 20min intervals until pain control achieved
- Pethidine is not used at St George’s Hospital any longer for sickle crises - it is a cerebral irritant which can cause seizures and has poor bioavailability
- If patients from elsewhere request pethidine please discuss with haematology SpR
- Morphine alternatives include oxycodone and hydromorphone
- Entonox should not be used after leaving the ambulance due to risk of irreversible neuropathy
- Adjuvant analgesics include paracetamol and NSAIDs (as long as no evidence of nephropathy)
- Please ensure laxatives, antiemetics and antipruritics are co-prescribed
- Ensure regular review of SpO₂s and respiratory rate in patients needing opioid analgesia

Supplementary management

- oxygen, keeping oxygen saturations above 94%
- iv fluids if not orally maintaining adequate hydration
- aim for 1 – 1.5 x maintenance volume once volume depletion corrected
- Broad spectrum antibiotics if signs of infection (Co-amoxiclav with clarithromycin or levofloxacin alone if penicillin allergic. (People with sickle cell disease are effectively asplenic and therefore susceptible to infection with encapsulated organisms such as Streptococcus pneumonia and Haemophilus influenzae B). Antibiotics can be given orally unless clinical concern warrants intravenous administration.
- Liaise with haematology SpR and if in hours the Sickle Cell Clinical Nurse Specialist for admissions
Medical assessment for complications requiring specific/urgent intervention and treatment (see below for details of life threatening crises)

- Full history and examination (focussing on chest, abdomen and CNS)
- Regular assessment of vital signs
- Blood samples for FBC, reticulocyte count, renal and liver profile, group and save
- Blood cultures if suspicion of infection
- CXR if chest signs / symptoms
- There is usually no need to XR painful bones in a simple pain crisis
- Blood transfusions are usually not indicated and should only be considered after discussion with haematology SpR

**Admission**

If the patient is to be admitted (most cases) immediately contact the Bed Manager and advise the Haematology team. No patient admitted with sickle cell crisis should be placed on a ward outside the Medical Service Centre - there are specific cohorted sickle beds. After admission to the ward continue 2 hourly sc morphine or analgesia regimen prescribed. Give at the dosage indicated on the patient’s personal protocol if available, with additional 5–10mg boluses for breakthrough pain. The patient should wait no more than 4 hours in A&E. During this wait and if delayed longer please ensure that:

1. the analgesia regimen is followed
2. the patient has fluid input maintained
3. the patient has antibiotic regimen maintained
4. the patient is observed regularly to ensure all vital signs are maintained and pain levels assessed.

*If a patient is discharged from, or leaves A&E, then:*

- contact the haemoglobinopathy specialist nurse (SGH blp 7520; or via Balham Health Centre on 0208 700 0615 if community-based) and give details of the admission and assessment.
- give the patient sufficient analgesia to ensure effective pain management until the patient may see their GP or a specialist nurse counsellor.

**LIFE-THREATENING CRISIS**

Patients can present with a variety of other acute manifestations which may be rapidly fatal if not recognised and treated quickly.

**Infection**

Patients prone to sickling have reduced splenic function and are at risk of overwhelming septicemia (pneumococcus, meningococcus, rarely haemophilus) even if taking penicillin prophylaxis. Peak risk is in childhood. The patient may present with fever, shock, seizures, coma, meningitis (often with delayed CSF pleocytosis) or even profuse diarrhoea.

Early IV antibiotics to cover pneumococcus and staphylococcus (Co-amoxiclav and clarithromycin, or if penicillin allergic then levofloxacin alone) and volume support are vital. If osteomyelitis suspected, discuss with Microbiology.

**Spleenic or Liver Sequestration**

During infection children may suffer a rapid fall in haemoglobin and growth of the spleen – changes often noted by the mother. Death can result from hypovolaemia and anaemia. Early transfusion is vital. In adults, liver sequestration is more common and can present similarly with profound anaemia and hepatomegaly. Transfusion is often required in these patients as well.
**Chest crisis**

Severe shunting & hypoxia caused by intra-pulmonary sickling and mimicking pulmonary embolus/pneumonia, may start in one lobe and then spread to others. It sometimes begins as a pain crisis affecting ribs or shoulders. Treat with fluids and oxygen; observe arterial O\(_2\) tensions – a falling PaO\(_2\) will require exchange transfusion and needs expert advice. Encourage patients with chest pain to attempt one maximal inhalation every 5-10 mins ('incentive spirometry') to aerate basal lung segments; this reduces the risk of progressive sickle chest syndrome. Non invasive respiratory support may well be required, as well as urgent exchange transfusion. Discuss with haematology urgently.

**Girdle syndrome**

If sickling occurs in the splanchic bed, abdominal pain with rigidity, loss of bowel sounds and increasing icterus may develop. IV fluids are vital. A surgeon should be consulted to exclude other abdominal events, but surgery should be withheld unless unavoidable, and then only after exchange transfusion and discussion with haematologists.

**Cerebral sickling**

Patients can present with strokes, fits, coma, bizarre behaviour or psychosis, and sickling should be excluded in any susceptible patient with such signs. IV fluids are vital and early exchange transfusion a possibility. Patients are at risk of both haemorrhagic and ischaemic strokes.

**Priapism**

Priapism typically affects only the corpora cavernosa. Major or prolonged attacks post puberty can result in permanent loss of erectile function. Urgent referral to Urology is essential as early decompression can be achieved by aspiration +/- intracavernosal phenylephrine.

**Blood transfusion**

In a patient with Sickle Cell Disease blood transfusion can be dangerous. Never give simple transfusion for anaemia (except in those sequestrating), without reducing HbS level by exchange. If this precaution is not taken the blood viscosity will increase and make the patient worse. Consider if Hb<5g/dl or if there has been a 2g/dl fall from steady state. Get haematological advice and ensure that the blood transfusion department knows that the patient due to receive blood has sickle cell, so that appropriately phenotyped blood can be provided.

**Surgery**

Do not plan or carry out surgery without first assessing the patient with the Haematology Team. Special pre- and post-operative care, often including blood exchange, is essential to optimise outcome.

**Acute renal failure**

In sickle cell disease this is most commonly multifactorial with causes including dehydration, sepsis, nephrotoxic drugs (especially NSAIDs) as well as acute papillary necrosis. Urine dipstick for haematuria is important, as is a MSU to exclude infection and these patients should be discussed with the nephrology team as well as haematology. Intravenous fluid replacement is important (minimum of 3 litres/24 hours) and ensure nephrotoxic drugs are withheld.