Sickle Cell Disease: Acute complications in Adults Guideline (including management of painful crisis) - 2409

EMSTN
East Midlands Sickle Cell and Thalassaemia Network

Guideline Number & Full Title:	*Sickle Cell Disease: Acute complications in Adults Guideline (including management of painful crisis) - 2409*
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Scope ***(Target audience, state if Trust wide):***	Clinical teams managing patients with Sickle Cell Disease across the trust.
Review date ***(when this version goes out of date):***	01/01/2027
Explicit definition of patient group to which it applies ***(e.g. inclusion and exclusion criteria, diagnosis):***	Adult patients with Sickle Cell Disease
Changes from previous version ***(not applicable if this is a new guideline, enter below if extensive):***	Guideline completely rewritten.
NICE guidance reference:	Overview | Sickle cell disease: managing acute painful episodes in hospital | Guidance | NICE https://www.nice.org.uk/guidance/cg143
Summary of evidence base this guideline has been created from: (other than NICE)	Sickle cell Society Standards for the clinical care of adults with sickle cell disease in the UK, February 2018
This guideline has been registered with the trust. However, clinical guidelines are guidelines only. The interpretation and application of clinical guidelines will remain the responsibility of the individual clinician. If in doubt contact a senior colleague or expert. Caution is advised when using guidelines after the review date or outside of the Trust.	

1. Introduction and who Guideline applies to

This guideline outlines the management of patients with sickle cell disease who are admitted acutely to hospital. Sickle cell disease (either HbSS or compound heterozygous states including HbSC and HbS/B-thalassaemia) affects around 15000 patients in the UK.

The commonest reason for admission to hospital is acute pain, which can occur in any part of the body but most often affects the limbs. Sickle pain is usually very severe and often requires opiate analgesia. Patients often have analgesia at home which they take before coming into hospital and can often verbalise if this is their 'usual' sickle pain.

The pathogenesis of sickle cell pain is due to deoxygenated sickle haemoglobin forming large polymers which cause red cells to become deformed and obstruct small blood vessels. Triggering factors include cold weather, dehydration, exam stress, pregnancy but often no cause is found.

These guidelines may be used by medical and nursing staff in any area of the hospital. Any admission of a patient with Sickle Cell Disease should be discussed with the Haematology team on call. They outline the care of patients admitted with acute complications of sickle cell disease.

2. Guideline Standards and Procedures

This guideline outlines the management of patients with sickle cell disease presenting with acute complications. As described, the commonest reason for admission is acute pain but other, potentially life-threatening complications are also discussed.

- The local policy for this trust is shown in ***Appendix 1***.
- A summary of this guideline document is available in ***Appendix 2***.

Arrangement for admission

- All patients known to the local haemoglobinopathy team will have been made aware of how to contact the clinical teams for advice, including out of hours. Each hospital will have their own system for ensuring open access for urgent medical review.

Immediate assessment

- Baseline observations must be performed as standard and NEWS documented. Escalations should be carried out in line with Local policy. If opiates have been administered by the ambulance crew, this MUST be documented clearly.

- Baseline oxygen saturations must be documented ON AIR. If <95%, ABG should be performed and more urgent escalation to haematology considered.

- Suspected sepsis should be managed in line with local policy

1. *Acute Painful episode*

1.1 Pain management

Acute painful sickle cell crises are caused by blockage of the small blood cells by sickled red blood cells. Painful episodes can occur unpredictably but may be precipitated by infection, dehydration, hypoxia or stress. Pain may fluctuate in intensity and frequency and patients will usually identify that their pain is typical for sickle pain. Patients will usually self-manage mild-moderate pain at home and will only present to hospital if they are unable to manage the pain with their usual analgesia. The patient is an expert in their condition and their views should be listened to and management discussed with them.

• A pain score MUST be documented. Analgesia should be addressed as a priority and in line with NICE quality standards, **analgesia must be administered within 30 minutes of admission (NICE CG143).**

• IV access is NOT mandated in all patients and delays for attempted cannulation are not an acceptable reason for not meeting this target.

• Entonox may be required until the first dose of analgesia is administered but must not be continued after opiate analgesia has been given.

• As part of the annual review process, individualised care plans may be available on careflow. If unavailable, patients should be managed as per the local painful crisis flowchart (See Appendix 1).
o Some complex patients with frequent admissions may also have a Individualised care plan available.

• If the pain is their 'usual sickle cell pain', patients will often have taken their own analgesia at home. This should be taken into consideration when prescribing analgesia. Opiate analgesia is most often required, providing baseline observations are stable. **Their pain should be assessed using the pain assessment tool.**

• Opiate naïve patients should be assessed based on their pain score and analgesia offered in line with the WHO analgesic ladder (below):

Step 1: mild pain (1-3)
Non-opioid ± adjuvant: Paracetamol 500mg-1000mg + Ibuprofen 400mg TDS
Step 2: moderate pain (4-6)
Weak opioid (or low dose of strong opioid) ± non-opioid ± adjuvant: As above for mild pain + Dihydrocodeine 30-60mg (maximum dose 180mg daily)
Step 3: severe pain (7-10)
Strong opioid ± non-opioid ± adjuvant As above for moderate pain + Morphine sulphate (see text)

- Morphine should be given via the most appropriate route. Some patients recently transitioned from paediatric care may prefer oral morphine and this should be offered. Unless otherwise documented, subcutaneous morphine should be given. Intravenous morphine is not recommended as routine.

• **If the patient does not have a dedicated analgesia plan, then give based on weight:**
o If ≤50kg, give 5mg morphine sulphate SC stat
o If >50kg, give 10mg morphine sulphate SC stat
Patients with tolerance may need a higher starting dose and a higher total dose.

Efficacy of analgesia should be re-assessed after **30 minutes** and a further dose of weight appropriate SC/IV morphine should be administered provided that there is no evidence of opioid toxicity.
• If the RR is <10 per minute, omit opioid analgesia
• If RR <6, give naloxone 100 microgram every 2 minutes as necessary

Pain should then be reassessed every 30 minutes until adequate pain control achieved.
If pain control is inadequate despite 2 doses of morphine:
- Discuss the case with the on call haematology registrar (mobile via switchboard) or local haemoglobinopathy nursing team (during working hours) and review patients specific care plan (if available)
- Consideration of escalation to patient controlled analgesia (see protocol)
- Consideration of alternative causes of pain if out of context with 'usual' sickle cell pain.
- Once pain is controlled, regular adjunct analgesia and PRN opiate analgesia (0.05- 0.1mcg/kg to nearest mg every 2 hours) should be prescribed.

Adjunct Analgesia	
Paracetamol 1gram IV/PO	Up to 4 gram in 24 hours *(providing weight > 50kg)*
Ibuprofen 400mg PO	Up to maximum dose of TDS unless contraindicated (such as patients with known renal issues/GI side effects)
Dihydrocodeine 30-60mg PO	Up to maximum dose of 180mg daily
Lidocaine 5% topical patches	Offer to all patients unless contraindicated
Non-pharmacological methods: Local heat packs TENS machine	May be available from haemoglobinopathy team if available

Alternative analgesics such as oxycodone, ketamine, methadone and fentanyl should only be used after discussion with the acute inpatient pain team and Consultant Haematologist unless specifically indicated in the patients care plan documentation.

If patients are on Modified release opioids (eg Modified Release Morphine Sulphate Tablets, (MST), Oxycodone or Fentanyl Patch), these can be continued.

Requirements for analgesia should be reconsidered on a daily basis by the haemoglobinopathy specialist team.

If there are difficulties controlling a patient's pain and further advice is required the inpatient pain team and Consultant Haematologist should be contacted.

Pethidine should not be used in any circumstance.

On discharge:

- Prescribe short acting opiates for no more than 5 days
- If starting Modified release opiates during admission these should be given for the shortest possible duration and a weaning plan should be written on the discharge letter.
- Sign post the patient to GP or follow up clinic for review of opiate prescribing if felt likely that ongoing opiates required.

1.2 Adjunct medication

Anti-pruritics:
•Prescribe chlorphenamine 4mg TDS/PRN PO or hydroxyzine 25mg PRN/BD

Anti-emetics:
•First line: Ondansetron 4-8mg BD IV/PO or Prochlorperazine 5-10mg TDS PO
•IV cyclizine use should be avoided (if requested and not in patients care plan this should be escalated to Sickle Cell Team/On Call Haematologist)

Laxatives:
•Ensure regular laxatives are prescribed if on regular morphine (Senna TT BD or Laxido 1-3 sachets daily). Patient's choice should be taken into consideration.

Stomach protection:
•Give concurrent PPI (Lansoprazole 30mg) if prescribed NSAIDs

Folic acid:
•Patients will be on regular folic acid 5mg OD long term, ensure this is prescribed.

Oxygen:

•There is no evidence for this being used routinely in all cases of painful crisis

•Its use should be dictated by the clinical situation and oxygen saturations

- O If sats <95% on air, perform ABG and give supplementary oxygen (titrated to maintain sats >98%)
- O If increasing oxygen is required, urgent medical review is required to assess for acute chest syndrome
- Many patients have a symptomatic benefit from Oxygen therapy, and it should be prescribed and be available whatever the oxygen saturations (even if >98%) if the patient requests.
- If Oxygen saturations on air <95%

 Call medical staff for urgent review

 Administer humidified oxygen at 2-4 L/min by mask or nasal cannulae

 Increase frequency of observations to hourly or more frequently if clinical picture dictates

 Check Arterial Blood Gases (ABGs) **on air**

 Blood tests, Chest X-Ray and other investigations as indicated by clinical picture

 Refer to ACUTE CHEST SYNDROME guidance.

1.3 Baseline Investigations

•Blood tests: FBC, Reticulocytes, Biochemistry (U&Es, LFTs, LDH, Bone, CRP), G&S

•Reasonable attempts should be made to perform baseline investigations in admission area. If unsuccessful due to difficult venous access this should be deferred until the patient is admitted to haematology **unless more urgent results are clinically indicated.**

•Haemoglobin electrophoresis only indicated if a new patient or patient recently transfused

- O If recently transfused (within 12 weeks), also request DAT/G&S (via blood bank)

•If appropriate: Blood cultures, Urine dipstick +/- MSU, Throat swabs (if viral symptoms), viral serology, COVID-19 swab

•CXR **not** routinely required: perform if dyspnoea, O2 sats<95%, increased respiratory rate, cough, fever or chest pain

1.4 Other considerations

Hydration:

- Adequate fluid intake is essential.
- Patients should be encouraged to drink at least 3 litres of water-based fluids per 24 hours.
- Patients requiring admission for a sickle crisis and who cannot take oral fluids need proper hydration evaluation and careful management.
- Assess hydration status: calculate according to degree of dehydration plus maintenance fluids.
- Not all patients require IV fluids but all patients should be on a fluid balance chart and supplementary fluids given if necessary

Antibiotics:

- Most patients will be on long term prophylaxis with Penicillin V 250mg BD. If admitted and therapeutic antibiotics are not required, this should continue
- If the patient is febrile or has a history suggestive of an infective cause of the sickle cell crisis they should be commenced on antibiotics in line with the Local microbiology guidance
- If patient is on Hydroxycarbamide (Hydroxyurea), check fbc urgently and stop the Hydroxycarbamide if the platelet count $<80x10^9$/l, reticulocytes $<80x10^9$/l or neutrophils $<1x10^9$/l

Thromboprophylaxis:

- Sickle cell disorders are associated with an increased risk of thrombotic complications
- All patients should be VTE assessed on admission and offered LMWH prophylaxis, unless contraindicated
- Anti-embolic stockings should **not** be used

Transfusion:

- See local transfusion guidelines

Patients with sickle cell disease often have a chronic anaemia of 60-100 g/L which is normal for them. Hb S has a lower oxygen affinity than Hb A so tissue oxygenation is better than expected. It is useful to check the patient's steady state Hb level (eg from an OPD appointment) when reviewing their blood count. The Hb may fall 10-20 g/L during a sickle crisis but blood transfusion is NOT routinely indicated and in fact may exacerbate a crisis.

- Any transfusion in sickle cell patients, other than for life threatening haemorrhage, should be discussed with the haematology team.
- Exchange transfusion may be required for complications including chest crisis, stroke and multi-organ failure: see *Sickle Cell Disease: Adult Transfusion Guideline.*

Ongoing patient monitoring:

- Observations should be carried out every half hour until pain is controlled.
- Respiratory rate, sedation score and O2 saturation should be checked 1-2 hourly during first 6 hours while on opiate analgesia. GCS and pupil size should also be noted.
- Temperature, BP and pulse, and fluid balance should be reviewed at minimum of 4 hourly.

Nice Guidance: 'Clinical assessment (clinical observations including pain, sedation, vitals signs, respiratory rate and oxygen saturations) should be performed hourly for the first six hours after admission or after dose escalation and at least four hourly whilst on opiates until discharge'.

Other acute presentations

2. Acute Chest Syndrome (ACS)

This is a life-threatening complication of sickle cell disease and can arise during a painful crisis or occur on admission (ACS is preceded by a 24-72hour history of VOC in 50% of cases).

Clinical Features - some, but not all, of the following may be present:

Chest Pain	Hypoxia (sats <95% or >3% below baseline)	Wheeze
Cough	Tachypnoea	Fever
Shortness of breath	Tachycardia	Fall in Haemoglobin

Please note physical signs often precede x-ray changes.
• crepitations • bronchial breath sounds • reduced air entry • dullness to percussion
• rhonchi

Diagnosis and Investigations
Based on:

- Clinical suspicion (low sats, hypoxia on ABG (performed on ***air***), new chest signs)
- CXR: new bilateral pulmonary infiltrates are typical (can lag behind clinical picture)
- Perform baseline investigations (as above)
- Microbiology: blood and sputum culture, atypical serology, viral swabs, urine legionella and pneumococcal antigen
- Patients with pre-existing respiratory disease at higher risk

Differential Diagnosis:
Acute Infection (including COVID-19 infection)
Pulmonary Embolism
Opiate Toxicity
Fluid overload
Hypoventilation due to pain
Asthma exacerbation

Assess severity

- Increased oxygen requirement
- Increase RR
- Decreasing platelet count
- Fall in Hb from baseline >10g/L
- Neurological symptoms
- Multi-lobar involvement on CXR

Management

- Preventative measures
  O Vigilance
  O Incentive spirometry for all patient if chest/rib pain on admission (and use as adjunct to treatment)
  O Early treatment of pain and infection with careful monitoring for opioid toxicity
  O Careful hydration management to avoid fluid overload

- Intervention if ACS suspected:
  O Inform haematology registrar/consultant (if not already aware)
  O Early liaison with CCD/outreach team as transfer may be necessary
  O Start appropriate antibiotics to include atypical cover
  O Chest physiotherapy
  O Nebulised bronchodilators may be useful
  O Oxygen to ensure sats maintained >95%
  O Consider PE- Can co-exist or precipitate chest crisis.
  Usually more sudden onset and patient recognises pain not typical of sickle cell pain. Consider CTPA/VQ scan if clinical suspicion of PE.

- Early transfusion can be lifesaving and should be discussed with a senior haematologist as soon as Chest Crisis suspected (SpR or Consultant)):
  O Simple top up may be adequate, especially if Hb decreased from baseline (post transfusion Hb <100g/L)
  O Manual or automated exchange transfusion preferable in a deteriorating patient or those at risk of hyperviscosity due to baseline Hb
  (see *Sickle Cell Disease: Adult Transfusion guideline*)
- Refer for assesessment by outreach +/- CCD and consideration of NIV if:
  □ pO2 (on air) <8.0kPa
  □ pCO2 >6.0kPa
  □ Rapid deterioration

On discharge:

- Ensure on appropriate abx prophylaxis
- Update vaccinations
- Offer hydroxycarbamide if not successful or not acceptable consider regular exchange programme to prevent recurrence of ACS.
- Request Outpatient sleep study and PFT's.

3. Acute Anaemia

- Presentation with pallor or extreme lethargy
- Investigations: FBC and reticulocytes, parvovirus serology, urgent cross match, DAT
- Consider underlying pathology including parvovirus infection, sequestration, folate deficiency or acute oxidative haemolysis if known G6PD deficiency. Screen for DHTR/Hyperhaemolysis if recent transfusion (see *Sickle Cell Disease: Adult Transfusion guideline*).
- Check parvovirus PCR if serology negative and high clinical suspicion.

4. Acute Neurological issues

4.1 Acute Stroke

- Stroke is a common complication of SCD and one of the leading causes of death. It can present with the typical features of stroke:
  o Limb weakness
  o Paraesthesia
  o Fits
  o Acute confusion

- *Acute ischaemic stroke* may occur at all ages, but most common in children and >30 years
  □ Precipitating factors: dehydration, fever
  □ Sometimes in otherwise well patients

- *Acute haemorrhagic stroke* (includes intracerebral, intraventricular and subarachnoid haemorrhage secondary to ruptured aneurysms and moya moya formations)
  □ Occurs at all ages, but median age of onset is 22 years
  □ Clinical presentation includes headache, loss of consciousness, hemiparesis, fits
  □ Often occur in context of acute illness (eg. sepsis)

Management:

- Urgent imaging: Patients should be assessed and initially managed in line with the local acute stroke guideline with urgent CT head and CT angiogram
- Clinical stabilisation: Keep warm, hydrated and oxygen therapy.
- Thrombolysis: should be considered in sickle cell patients with acute cerebral thrombosis who otherwise meet current UK national recommendations, but these cases should be discussed with a senior haematologist and stroke physician
- Transfusion: Urgent exchange transfusion should be performed to reduce HbS to <20%. Initial top up transfusion can be considered if Hb <60g/L. The aim is to improve oxygenation and minimise damage occurred. Acutely, this may need to be performed as a manual exchange with an automated exchange at the earliest opportunity. Automated exchange is preferred due to less lability of blood pressure (please note can lead to transient thrombocytopenia).
- Haemorrhagic stroke should be managed as per neurosurgical advice (exchange transfusion may need to happen post-surgery if this is required)
- Following acute intervention, an MRI/A head should be performed to assess for sickle related vasculopathy and determine long term management
- Other causes of stroke seen in young adults without sickle cell disease should also be considered (thrombophilia, CNS infection, illicit drugs, arterial dissection, congenital heart disease)

4.2 Headache

- The first episode of acute severe headache or a significant change in type of headache should be evaluated as an emergency and the diagnoses of intracranial haemorrhage or venous sinus thrombosis considered.
- Discussion with the neurology team and appropriate imaging should be considered.

4.3 Migraine

- If acute or prolonged migraine attack suspected, discuss with neurology team.
- Consider treatment with high dose NSAIDs (oral if tolerated: eg. 900mg Dispersible aspirin or 800mg Ibuprofen or 750mg Naproxen), prokinetic antiemetics (eg. Domperidone or Metoclopramide) if able to tolerate orally.
- If significant vomiting, treat with IV fluids (with supplemental IV magnesium, eg 1g in 1L saline), IV NSAIDs (eg IV Ketoprofen) and/or IM Prochlorperazine.
- Consider a triptan if within the first 6-12 hours of an attack
- Chronic headache (>15 headache days a month), is common in SCD. Chronic migraine is a common cause of headaches in patients with SCD.

4.4 Other neurological complications

- Fits – treat as in non-sickle cell population. Screen for acute ischaemic stroke or haemorrhage.
- Venous sinus thrombosis – may have an increased complication rate in SCD. Diagnosis made by MR Venogram and treated with anti-coagulation
- Posterior reversible encephalopathy syndrome (PRES) – seen typically in context of child with severe ACS and occasionally in adults and in pregnancy. Characterized by neurological deterioration, headache and seizures. Diagnosis made by MRI and treatment is supportive.
- Silent cerebral infarction – these are common in children and adults with SCD. They are characterized by MRI lesions without overt neurological impairment and are associated with cognitive impairment.

5. Abdominal Pain and Jaundice

Patients can present with abdominal pain for a number of reasons. The differential diagnosis includes (not exhaustive list):

Gallstones	Referred pain	Acute splenic/hepatic sequestration
Pancreatitis	Appendicitis or other infection	Constipation
Bowel Ischaemia	Thrombosis i.e. renal vein	Vaso-occlusion
Peptic ulcer disease	Ischaemic colitis	Biliary issues

Each patient with Sickle Cell Disease has a baseline degree of hyperbilirubinaemia. Elevations in bilirubin levels can result from:

- Haemolysis which may be exacerbated during a vaso-occlusive crisis, delayed transfusion reaction and hyperhaemolysis
- Obstructive causes
- Intrahepatic cholestasis

Investigations:

- FBC, retics, U&Es, LFTs, Coagulation, Lipase, Blood cultures, MSU
- Relevant imaging, as guided by clinical features/findings and radiology advice Take radiation exposure for patients with frequent admissions into consideration

5.1 Acute Sequestration

- Sudden enlargement of the spleen (or more rarely, liver) in which blood is pooled in the organ leading to a severe reduction in circulating red cells and profound anaemia
- Abdominal examination will reveal an enlarged spleen or liver
- Marked anaemia with reticulocytosis and conjugated hyperbilirubinaemia
- Urgent FBC and retics followed by urgent cross match as a top up transfusion may be required. See *Sickle Cell Disease Adult Transfusion guideline*.
- There may be co-existing infections including salmonella species therefore consider broad spectrum antibiotics.

5.2 Gall Stones

Demonstration of gall stones in a patient with abdominal pain does not mean that the gall stones are the cause of the pain.

- Occur in at least 30% of children and over 70% of adults
- Often asymptomatic but can cause: Acute cholecystitis

Other complications include

- Chronic cholecystitis
- Biliary colic
- Obstruction of the common bile duct with acute pancreatitis
- Can precipitate abdominal painful crises and the mesenteric syndrome

Biliary sludge may be a precursor to gall stone formation

Diagnosis:

- Plain abdominal X-ray (as many as 50% of stones may be radio-opaque)
- Ultrasound

Differential diagnosis:

- Hepatitis (viral)
- Peptic ulcer
- Vaso-occlusive episodes
- Hepatic sequestration
- Chest syndrome

Management:
Acute episode:

- Antispasmodics
- Hydration
- If acute cholecystitis/cholangitis is suspected prescribe antibiotics :
  As per hospital guidelines for biliary sepsis
- Avoid pethidine in view of risk of fits

Common bile duct obstruction:

- Surgical review for MRCP or Endoscopic retrograde cholangiopancreatography (ERCP) or emergency surgery
- Once acute episode has settled: Elective cholecystectomy; generally laparoscopic is recommended

5.3 Abdominal crisis (Girdle syndrome or Mesenteric syndrome)

- Patients may present with mild abdominal pain in the context of an acute painful crisis.
- With abdominal crisis of mild-moderate severity the abdomen may be soft with mild tenderness but if severe examination may show a silent, distended abdomen without localising signs or rebound
- Some hepatic enlargement and preceding pain in back, abdomen, or limbs is common
- Often associated with bilateral basal lung consolidation
- Characteristic distended bowel loops or fluid levels on X-ray

Management:

- Serum lipase
- Oxygen, hydration and analgesia
- Nasogastric suction. Sips by mouth only
- Surgical intervention is not usually required but other causes of abdominal pain should be considered and surgical review is usually required

5.4 Sickle hepatopathy (Intrahepatic cholestasis):

Some patients experience episodes of severe hyperbilirubinaemia, associated with fever and hepatic pain in the absence of demonstrable stones. These episodes are thought to be due to severe intrahepatic sickling (intra-sinusoidal sickling and intracanalicular cholestasis).

Presents with:

- Severe right upper quadrant, acute hepatomegaly, coagulopathy, extreme hyperbilirubinaemia (mainly conjugated) with moderately elevated liver enzymes.
- Risk of progression to acute hepatic failure

Management:

- Exclude other causes of liver dysfunction including US abdomen and MRCP to exclude cholestasis.
- Liver biopsy relatively contraindicated in hepatopathy due to SCD due to risk of bleeding and other complications
- Early exchange transfusion to target S% of 20-30% can reduce mortality.

5.5 Splenic infarction

- This can occur in patients with redisdual spleens, most commonly in patients with Hb SC disease.
- Often identified on US abdomen.
- Treatment is supportive

6. Priapism - See Sickle Cell Disease: Priapism Guideline

Defined as a prolonged penile erection which is maintained without sexual stimulation and persists despite ejaculation and orgasm.
There are two types:
Stuttering (recurrent episodes lasting <30mins)
Ischaemic fulminant (>3 hours) – should be treated as a medical emergency.

Management

- Treat as Vaso-occlusive crisis: Hydration, warmth, analgesia, oxygen
- Encourage micturition
- Etilefrine 50mg may be given by mouth (available from main pharmacy)
- Urgent referral to on call urology registrar
- if persists > 3-4 hours, then aspiration of corpora cavernosa (with or without irrigation) maybe required with the instillation of adrenergic agonist e.g phenylephrine, progressing to a surgical shunt procedure if not successful
- Transfusion therapy is not thought to be useful in acute presentation although it may be considered if all other treatment is ineffective

7. Sepsis – including osteomyelitis

•Patients with sickle cell disease are immunocompromised and any features of infection should be managed in line with the local sepsis policy (***ensure sepsis six completed***)

•Blood cultures should be sent in any febrile patient

•Microbiology advice should be sought for suspected cases to ensure adequate pathogen cover in line with local policy

Patients with SCD are hyposplenic (auto-infarction of spleen) and at increased risk of infection particularly with (Haemophilus influenzae, Strep pneumoniae and Neisseria meningitidis).

Therefore consider:

 Pneumococcal sepsis (especially if not taking prophylaxis and not vaccinated). Penicillin resistant pneumococcus has been seen in the UK.

 Gram negative sepsis

 Lower respiratory tract infection (especially Influenza: perform viral swab and isolate patient)

 Urinary tract infection

 Osteomyelitis

 Malaria and Ebola if travelled recently

 Parvovirus B19- transient red cell aplasia- if low Hb and low reticulocyte count

 Yersinia if on desferrioxamine and have diarrhoea – contact infection team for advice

7.1 Osteomyelitis

•Osteomyelitis can be difficult to distinguish from vaso-occlusion and should be considered in any patients with a hot and painful limb.

•Both can present with pain, localised tenderness, warmth, swelling, fever and leucocytosis.

•Commonest causes: Salmonella (especially the non-typical serotypes such as Salmonella typhimurium, Salmonella enteritidis), followed by Staphylococcus aureus and gram negative enteric bacilli

Investigation:

•Take blood cultures if Osteomyelitis suspected

•Involve Orthopaedics to consider early aspiration/bone biopsy

•XR should be performed, but may be normal and MRI is likely to be required

Treatment:

•Initially broad-spectrum antibiotics to cover Salmonella and Staphylococcus pending culture results. Liaise with microbiology if systemically well consider holding antibiotics until microbiological samples collected.

•Continue antibiotics for minimum of 6 weeks.

•Consider drainage for fluid accumulation that does not respond to antibiotics.

8. Renal complications

•Patients with sickle disease are at increased risk of nephropathy and renal impairment. U&Es should be performed as part of baseline investigations for all patients presenting to hospital.

•Haematuria may occur and can be associated with papillary necrosis. This may be associated with ureteric colic and can lead to significant blood loss. Treatment is supportive and urological support may be required for renal irrigation.

•Other causes of renal impairment and haematuria should also be considered.

AVOID NSAID'S IN ALL PATIENTS WITH KNOWN RENAL IMPAIRMENT

8.1 Acute kidney injury (AKI)

Acute kidney injury (AKI) can be precipitated by dehydration, sepsis, drugs or in the context of multi-organ failure or on background of chronic renal failure and can occur in over 10% of SCD patients admitted to hospital.

 Assess renal function in all SCD patients admitted acutely to hospital

 Monitor fluid balance throughout hospital admission

In patients with AKI

 Exclude reversible cause

 Ensure aggressive fluid replacement

 Good blood pressure control and oxygenation

 Exclude/treat sepsis

 Renal USS to exclude post-renal cause

 Renal replacement therapy as indicated

 Hyperkalaemia is common (due to tubular defects) and needs careful monitoring

8.2 Haematuria

Microscopic haematuria is common in sickle cell disease due to microscopic infarcts in the kidney.

Frank haematuria is also common and is often due to renal papillary necrosis. This can be profuse. It may also occur in patients with sickle cell trait. It is usually painful and passing of renal papillae can cause renal colic and ureteric blockage.

In patients >40 years or with painless haematuria, alternative diagnoses should be considered (eg bladder tumours) and they should be referred to Urology.

Investigations:

 Ultrasound scan is the first line of investigation but hydrated intravenous urography may be necessary to establish the diagnosis.

Management:

 FBC, renal function and Group and Screen/Cross match

 Hydration (iv fluids)

 Treat infection

 Contact Haematology and refer to urology

 Avoid NSAID's

NB: Patients with delayed haemolytic transfusion reaction or hyperhaemolysis can present with 'coca cola' coloured urine. Therefore always ask for history of recent transfusion (refer to *Sickle Cell Disease: Adult Transfusion Guideline*)

8.3 Urinary tract infections

Common, particularly in women with SCD (especially during pregnancy). Should be vigorously treated to prevent serious renal pathology. Haematuria, secondary to papillary necrosis, can precipitate UTI, but other factors must be excluded.

9. Acute Visual loss

•Sickle cell disease is associated with retinopathy and any visual loss should be considered a medical emergency
•Patients should be advised to attend local eye casualty at their local hospital for urgent review.
•Considerations should be given to a central nervous event (e.g. occipital stroke) in patients with loss of vision but normal ophthalmological investigations
•Retinal artery occlusions should be managed as a stroke.

3. Monitoring compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
NICE pain score audit data	Regularly submitted to NHS England haemoglobinopathy dashboard and reviewed at East Midlands haemoglobinopathy network	Matt Player	Quarterly	Via dashboard

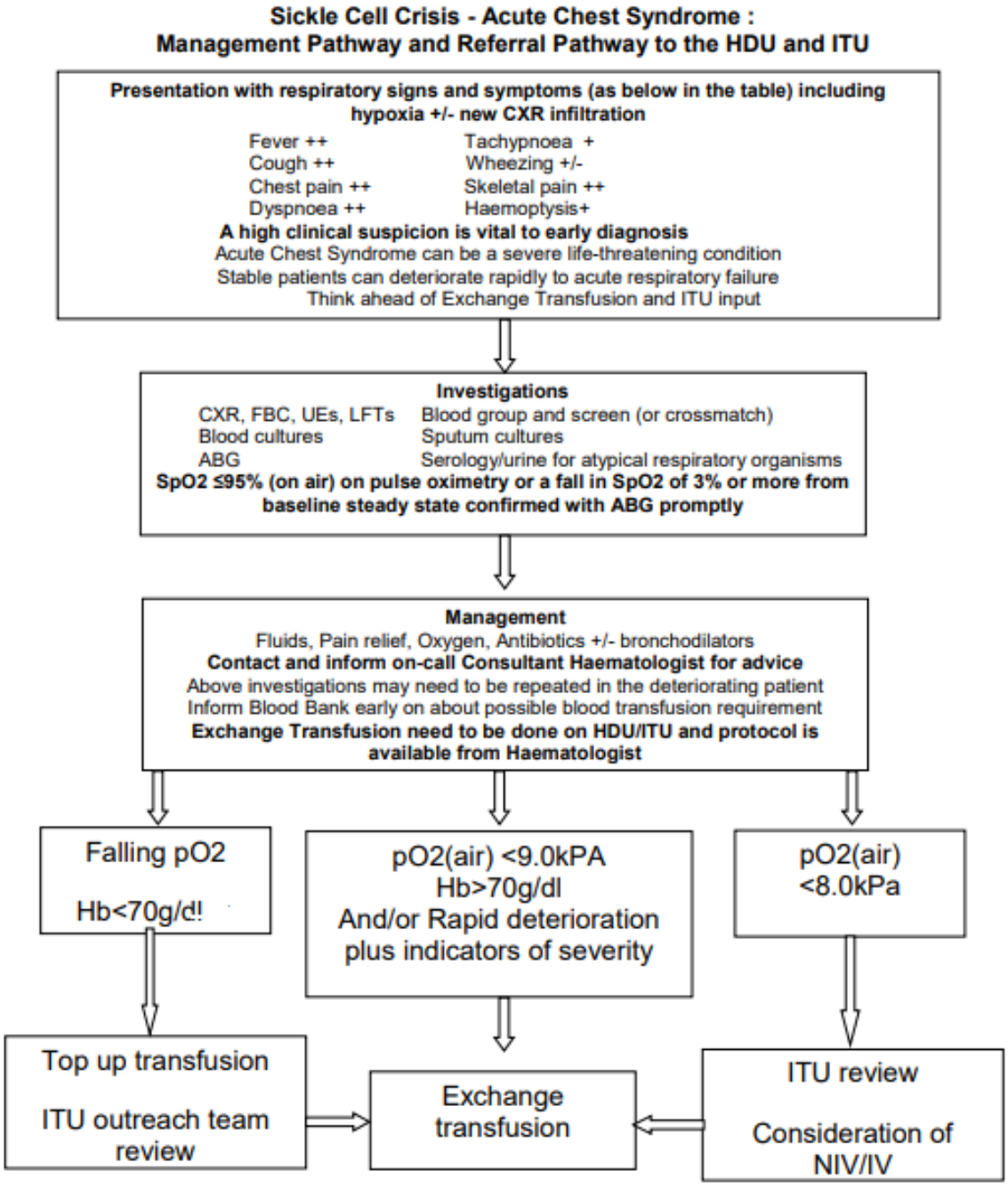
4. Supporting References

1. Standards for the clinical care of adults with sickle cell disease in the UK, February 2018. Sickle Cell Society.
2. Guideline on the management of acute complications of Sickle Cell Disease. West Midlands Sickle Cell and Thalassaemia Network.
3. Acute Complications of Sickle Cell Disease. St Georges Hospital.

Appendix 1: Sickle Cell Disease – Acute Crisis Guidance for first 24 hours

Final Acute painful crisis pathway flowcha

Appendix 2- Summary of Management of Acute Chest Syndrome



Appendix 3: Acute Presentation Summary

Acute Presentation
Guideline summary a

Appendix 4: Nursing Care plan- Management of a Patient Admitted With a Sickle Crisis

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Management of a Pati